COLOR ATLAS AND MANAGEMENT OF
VASCULAR DISEASE

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VASCULAR DISEASE

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PREFACE

Vascular diseases first received serious clinical attention in this country about 30 years ago In the ensuing period there have been great advances in diagnosis and therapy These have been subjected by the authors to constant testing in their hospital services and practices The authors have also worked with the American Medical Association the American Heart Association and the New York Heart Association to establish nationwide criteria for nomenclature labora tory findings and clinical applications pertinent to the problems of circulatory disease

There has been a great deal of literature on the subject but there has been no book of color description Such a book will be most helpful to the practicing physician who has to deal with problems of diagnosis The color changes which must be understood for accurate diagnosis cannot be portrayed by black and white illustrations The *Color Atlas and Management of Vascular Disease* supplies this need From files including thousands of case histories carefully selected cases are discussed and the diagnosis and pathogenesis of the diseases are explained with their treatment The principles of therapy are on the whole conservative but some cases are described in which radical measures were effec tive The details of surgical technic are not included because they are beyond the scope of this *Atlas* Nevertheless surgeons will be greatly interested in the conditions illustrated

The pictures presented in this *Color Atlas* were chosen from a collection of 600 photographs illustrating 200 cases started in 1946 These 192 color photographs illustrate the lesions of 94 patients which were taken by us or by Edgar J Nebel of the Clay Adams Company These are also reproduced in transparency form by the Clay Adams Company and duplicate slides may be obtained from them for educational purposes or for illustrated lectures

Nomenclature and Criteria for Diagnosis of the Diseases of the Heart and Blood Vessels published by the New York Heart Association in 1953 has been used as the basis for terminology We should like to thank Dr Harold Pardee chairman of the parent committee for his permission to draw liberally from the chapter on peripheral vascular disease in writing this *Atlas* We should also thank our associates on the subcommittee Dr Nelson W Barker Dr Paul J Klemperer Dr Joseph E Flynn Dr Gerald H Pratt and Dr Wallace M Yater who helped prepare this section of the nomenclature and criteria

Miss Esther Judkins and Miss Daphne Morse of the library of The Rockefeller

Institute and Miss Ann F Burke of the library of the Cornell University Medical College, have helped with the preparation of the bibliography

The staff of the Vascular Clinic at Cornell including Dr Ellen McDevitt Dr Robert Huebner, Dr Francis Perrone Dr Jerrold Lieberman Dr Clara Gross Dr Irwin Werner Dr Walter Palmer, and Dr Leonard Schuyler have participated in the care of these patients

The final manuscript was typed by Mrs Barbara W Gatje of our research staff Earlier versions were edited typed and proofread by Miss Gertrude C Smith and Miss Gertrude Rogers of The Rockefeller Institute Miss Gerd Stokke and Miss Irene R Moullin assisted with the galleys We thank each of them for *their enthusiastic and expert help*

The cost of color printing is so great that it would not have been possible to publish this book without the aid of Dr and Mrs Samuel Milbank General John Morgan Mrs Frank Corbett Mr Thornton Oxnard Mr Jean Lambert Drs Eugene Daly and Paul MacLeod of the Eaton Laboratories and the late Mr Isaac Harter who have contributed toward the cost of production

WILLIAM T FOLEY
IRVING S WRIGHT

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COLOR ATLAS AND MANAGEMENT OF
VASCULAR DISEASE

1

Arterial Diseases

ACUTE ARTERIAL OCCLUSION

Arterial occlusion may be caused by many pathologic states However the most common causes are thrombosis over the site of an atherosclerotic plaque embolization from a fibrillating or infarcted heart and thrombosis associated with the inflammation of thromboangiitis obliterans

Diagnostic Signs The diagnostic signs of acute arterial occlusion in a limb are pallor on elevation cyanosis at a horizontal or dependent level in the early stages coolness of the affected area absent or greatly diminished peripheral arterial pulses and subnormal oscillometric readings Pain and numbness are usually present The oscillometric readings are of value in indicating the level at which the obstruction occurs Pulses which are occluded or diminished by vasospasm may be released from this spasm and restored by sublingual nitroglycerin given as a diagnostic test (1)

Physiologic Changes (Diagram 1) Following the formation of the clot or the lodgement of the embolus a powerful vasoconstrictor agent appears to be liberated from the thrombus This has been named serotonin" by Rapport Green and Page (2) It produces a spasm of the affected vessel and the adjacent collaterals This spasm may extend distally for many centimeters but may relax over a period of hours or days after the first impact

The thrombus has a tendency to propagate itself distally in the direction of the decreasing pressure gradient and also in many cases centrally Blood flow through the spastic collateral vessels becomes very sluggish There is a tendency for sluggish blood to clot in both arteries and veins and in the capillary beds (thrombosis in situ)

Adaptive Mechanisms To counterbalance the arterial occlusion and vasospasm of collaterals several compensating mechanisms are developed Von Recklinghausen (3) in 1883 pointed out that the blood pressure rose above an area of arterial blockage and fell below this site This produces a pressure gradient which promotes the development and efficiency of collateral flow

Bier (4) in 1897 demonstrated that in an area with reduced arterial blood flow catabolic products accumulate These products have a profound vasodilator or histamine like action They tend to overcome the vasoconstrictor action of serotonin and result in a marked dilatation of collateral vessels

Thoma (5) in 1884 showed that the growth of collateral channels is proportional to the blood flow through them Lewis (6) in 1940 in a review of all the experimental data concerning collateral flow concluded that "the essence of the matter seems to be that there is a local call by tissues in need and that to this call there is a local and adequate response"

and extending to include the iliac arteries and segments of the femoral arteries have been successfully used They find their greatest value in the resection of aneurysms athero sclerotic occlusion as with the Leriche syn drome and vessels infiltrated with tumor The vessel above and below and especially at the site of attachment of the graft must be healthy and patent for this procedure to be successful It has been limited by the diffi culty in obtaining suitable homografts The development of plastic materials may over come this problem

2 The Use of Gravity To aid the flow of blood into collateral channels the affected limb should be kept below heart level This may be accomplished by elevating the head of the bed on six to eight inch blocks The common error of raising the involved limb on a pillow is to be deplored Alternately filling the limb with blood by making it dependent and then draining it by elevating it slightly can be conveniently accomplished by the oscil lating bed of which several types are avail able The bed must be regulated to suit each case The aim should be to obtain just enough elevation on the upswing to drain the veins but not to maintain it long enough to produce marked pallor The downswing should give ample gravity pressure flow to produce rubor if possible without reaching an angle uncom fortable to the patient

3 Vasodilation Collateral vessels are very ensitive to environmental changes both iemical and physical Slight chilling can ause extreme vasoconstriction Proper warmth uces dilation of the arterial tree

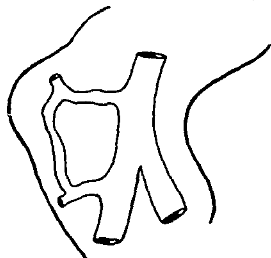
(a) Avoidance of Tobacco The use of obacco in any form produces profound vaso onstriction Some patients are much more ensitive to this than others In these even a ingle puff of a cigarette may cause measur able vasospasm Complete abstinence from obacco is a sine qua non for successful ther apy In addition to acting as a vasoconstricting agent in thromboangiitis obliterans there is evidence to show that the inflammation which is the characteristic pathogenic feature of the disease may be due to a peculiar idiosyncrasy to tobacco The inflammation is markedly ag gravated by the use of tobacco

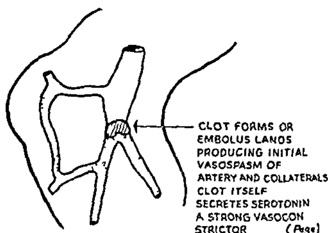
(b) Warmth Heat should never be ap plied to an area with decreased arterial flow However when the area proximal to it is warmed a reflex vasodilatation may follow For example if the popliteal artery is oc cluded a heating pad may be placed on the abdomen or groin with benefit

(c) Drugs

(1) *Systemic* The use of general vaso dilators such as alcohol nicotinic acid deriva tives hexamethonium compounds priscoline regitine dibenzoline tetraethyl ammonium halides ilidar arlidin etc has the disad vantage of dilating the minute vascular bed in normal areas as well as in the affected area where this effect is desired The net result may be to decrease blood flow to the limb where it is most needed but where spasm due to pain makes the vessels most resistant to dilatation Some of these agents also open up the numerous arteriovenous shunts in the limb and divert blood flow from the peripheral tissues such as those of the toes or fingers to this by pass system further reducing flow where it is required In our hands sublingual glyceryl trinitrate in a dose of 0 0004 gm has given the maximum degree of dilatation of large and small arteries for periods of one to two hours It does not however affect the musculature of the arterioles Therefore ex cept in cases of occlusion by spasm of the major arteries one should not expect an eleva tion of the temperature of the skin by this drug

(2) *Intraarterial* Repeated injections into an artery traumatize it This is more serious when an irritating substance is injected and there is some local infiltration of the wall of the vessel and surrounding tissues Reflex





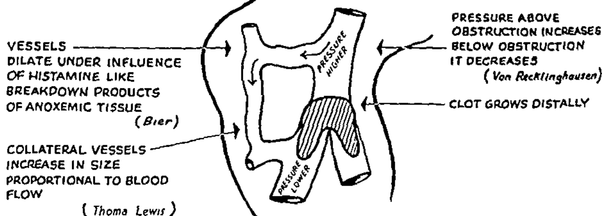


Diagram 1

TREATMENT

Successful management depends on the knowledge of the above physiologic data and the application of hygienic measures to aid the natural adaptive mechanisms Attention to minute detail is necessary (7)

1 Direct Surgery In the case of sudden embolization if expert vascular surgery is available if less than eight hours have transpired and if the general condition of the patient will tolerate it embolectomy is the treatment of choice This is practical only if the obstruction is at the bifurcation of the aorta or in the iliac artery or femoral artery down to the popliteal area Popliteal emboli and those distal to it do better without surgery Embolectomy may also be considered in the subclavian or axillary arteries It is now considered wise to recommend anticoagulant therapy during the postoperative course

The use of arterial grafts and by pass technics has made surgical intervention feasible in some cases Large grafts replacing the abdominal aorta distal to the renal arteries

dication is not produced Of course mobilization cannot be permitted during the stage when massive gangrene is threatened Spreading or very active infection also constitutes a contraindication As soon as this phase is passed we encourage carefully controlled and observed walking even when gangrenous toes or ulcers are present if a sharp line of demarcation has developed If there is indication of aggravation as a result of this it should be deferred until a later date

7 General Measures Supportive measures should not be neglected while focusing attention on a threatened limb Problems associated with serious underlying diseases such as polycythemia old rheumatic heart disease myocardial infarction or diabetes mellitus should be treated appropriately

CHRONIC ARTERIAL INSUFFICIENCY

If the limb survives the acute occlusion a state of chronic arterial insufficiency often follows Closure of an artery may also be very gradual The patient may then develop intermittent claudication necrotic ulcers or gangrene

TREATMENT

1 Exercise In the chronic stage exercise is important to encourage increased blood flow and develop collateral vessels Walking is most desirable but must be modified as to pace and distance by the capacity of the individual and his affected circulation Ideally it should be at a slow enough pace to avoid claudication pain or fatigue When this does develop the patient should pause and allow it to subside before resuming walking We have found that many patients who at first could walk only a few hundred yards or less have been able to increase their walking distance up to several miles This may of course be in part due to all other methods of therapy used concurrently These include

2 *The Use of Gravity* The patient should sleep with the head of his bed elevated six to eight inches or if possible use an oscillating bed The use of pillows for elevation is not satisfactory The elevation may best be achieved by blocks (chock type) under the posts at the head of the bed

3 Vasodilatation

(a) Avoidance of Tobacco *See above*

(b) Warmth In addition to reflex heat mentioned above if open lesions are not present the ambulant patient may take warm tub baths daily using 12 to 14 inches of water at 98 to 100° F (36 to 38 C) for 30 minutes

4 Drugs If skin lesions are present vasodilators which shunt the blood to the skin circulation may be of value to help heal these although it should be remembered that one may thus reduce the flow to the deeper tissues Alcoholic beverages are probably as effective as other means and well accepted by middle aged and older people To aid in dilatation of the large arteries the patient may sit or lie for an hour with a heating pad over the lower part of the abdomen At the same time he may take a tablet of glyceryl trinitrate 0 0004 gm under his tongue This procedure should be carried out one to three times daily

5 Anticoagulant Therapy Long term anticoagulant therapy is of special value in patients with rheumatic heart disease who have demonstrated any tendency to develop thromboembolic complications *In a group of such patients which we have followed* for from 1 to 11 years we have been able to reduce the *recurrence of such complications* by about 80 per cent (10)

6 *Sympathectomy* Sympathectomy is only of value in rare carefully selected cases It gives best results in the treatment of increased superficial vasomotor tone with *ulcerations* Increased blood flow to the muscles should not

spasm of severe degree is sometimes produced and gangrene may result Such procedures should therefore be reserved for tests of vital importance This therapeutic approach is rarely justified

(d) NERVE BLOCKS The most effective method for releasing vasospasm in the vessels of the skin and subcutaneous tissues of the legs is the injection of procaine into the lumbar sympathetic ganglia It frequently lasts for periods up to eight hours The results are apt to be less satisfactory when this technic is applied to the upper extremities Deep injections such as these should not be given if the patient is on anticoagulant therapy because of danger of hemorrhage This procedure also has the disadvantage of opening up arteriovenous shunts in the proximal segments of the limb thus reducing blood flow to the peripheral tissues

(e) SYMPATHECTOMY Rarely sympathectomy (8) may be justified in the treatment of acute occlusion It has the effect of prolonging sympathetic interruption but the disadvantage of subjecting an often severely ill patient to a major procedure

(f) INDUCED FEVER General vasodilatation occurs in fever This may be induced by intravenous injection of typhoid vaccine An initial dose of 5 million organisms using a dilution of 100 million organisms per ml is administered increasing by 3 to 5 million organisms with each dose unless the previous reaction induces a chill or fever of more than 3° C The aim is to produce a slight fever without a chill This form of therapy is most helpful in cases of thromboangiitis obliterans with marked vasospasm

4 **Maintenance of Tissue Viability** While the limb with major arterial insufficiency is awaiting the development of collateral flow the tissue in jeopardy must be protected against trauma maceration infection dehydration and thermal damage It is unwise to increase its metabolic rate and hence the demands for blood by the application of external heat The increased oxygen need cannot be met the deficits for oxygen and other tissue foods increase markedly and the tissues die On the other hand cooling produces vasoconstriction and decreased blood flow In general the affected limb should be kept in an environment of 70 to 88° F Wet compresses are contraindicated because of their tendency toward cooling and the heat loss by evaporation further chills the part Gently coating the limb with an inert fat combined with antimicrobial agents and then wrapping it loosely with cotton wool will help maintain an ideal environment and protect it against maceration

Cold or ice are used in selected patients only when it has been decided to amputate Such patients are typically diabetics in poor condition and with marked infection or toxic absorption from a gangrenous foot When used in this way they relieve pain reduce absorption from an infected area reduce fever and allow time for the patient to be prepared satisfactorily for the operation This frequently includes the proper control of a diabetic or cardiac condition

5 **Anticoagulants** In patients with embolization as for example from a heart in auricular fibrillation anticoagulants are useful in reducing the incidence of further emboli They also aid in the prevention of extension of thromboses During the period of greatly reduced blood flow in the affected limb thrombosis in situ in the capillary bed and small vessels together with thromboses in the veins is a constant threat which adequate anticoagulant administration will help combat

6 **Exercise** Under the heading of Adaptive Mechanisms it was pointed out that collateral vessels develop in size and efficiency proportional to the rate of blood flow One of the greatest stimuli to increased blood flow is the use of muscles Weight bearing should be encouraged as soon as the tissue viability will tolerate it (9) Patients should walk very slowly slow enough so that intermittent clau

CASE HISTORIES

ATHEROSCLEROSIS OBLITERANS

The gradual accumulation of atheromas in the intima of arteries leads to narrowing of these vessels Blood flow to the muscles may be sufficient when the person is at rest but when he walks it is not adequate Insufficient oxygen is furnished for muscular contraction Intermittent claudication results This is well illustrated by the following case history

Case 1 Intermittent Claudication due to Atherosclerosis Obliterans



Note intense rubor of affected leg on dependency



Note intense pallor of affected foot on elevation

A 60 year old railroad trainman had severe intermittent claudication in his left leg with color changes in the foot

In the left figure note the intense rubor that develops in the affected leg upon dependency

Intense pallor develops in the affected foot on elevation as shown in the right figure

These color changes are characteristic of arterial insufficiency Pulses and oscillometric readings were as follows

PULSES	RIGHT	LEFT
Posterior tibial	2+	0
Dorsalis pedis	2+	0
Femoral	2+	very faint
OSCILLOMETRIC READINGS		
Foot	1 0	0 0
Ankle	2 5	0 1
Calf	3 5	0 2
Thigh	3 5	0 4

Conservative therapy was instituted His rest pain ceased in two weeks His claudication distance has slowly improved and after six months he was able to walk up to one half mile at a moderate pace

As the disease progresses some vessels become completely blocked by atheromas In others a thrombus forms which suddenly interferes with the flow of blood When areas receive insufficient blood even at rest death of tissue results In the presence of diabetes mellitus this process takes place at a more rapid rate The small branch vessels as well as the large main trunks are frequently damaged and infection often complicates the picture

Case 2 illustrates these features

be expected from the operation In patients with normal or decreased vasomotor tone the results are usually disappointing Sympathectomy should be reserved for patients who have not done well on the conservative regimen outlined above including the absolute prohibition of tobacco The regimen is valueless unless the patient discontinues the use of tobacco

7 Care of Ulcers Necrotic ulcers must be cared for with great gentleness Debridement should be done minutely and at many sittings Amputation of necrotic toes often leaves a nonhealing stump Hot soaks must be strictly avoided for the reasons given above The temperature of saline soaks used daily should be from 90 to 95° F (32 to 35° C) Wet dressings are too cooling Strong chemicals only add to the necrosis Mild antimicrobial ointments with a wide spectrum are best such as aureomycin ointment 3 per cent Furacin soluble dressing is excellent if the physician is alert to skin sensitization These ointments have wide spectra are relatively nontoxic and kill many saprophytes as well

8 General Hygiene The patient must avoid chilling In cold weather he should wear heavy underwear and stadium boots He should be cautioned never to put heat in any form (including diathermy) on the involved limb (see paragraph 4 *under "Acute Occlusion*) He must avoid vasoconstrictor drugs such as epinephrine ephedrine benzedrine dexedrine or ergot derivatives He should always keep the affected limb below heart level

PROGNOSIS

The outlook for patients with chronic arterial occlusive disease is much better than commonly accepted by the medical profession provided that these patients persist in their adherence to the regimen outlined above This may mean a matter of years rather than weeks and the patient must be oriented regarding the long range approach to his problem at an early visit Many patients with major occlusions even of so large a vessel as the lower abdominal aorta have been restored to an active life and are in good health a decade or more later Some are able to walk long distances at a slow pace resume laborious activity and sports such as golf which do not require the running involved in tennis or squash

SUMMARY

If a sound knowledge of physiologic principles is applied nature will provide many adaptive measures to produce collateral blood flow Walking long continuous distances at a slow enough pace so that pain does not arise is most important Tobacco *must be eliminated Gravity should be utilized to aid* blood flow and elevation of the affected extremity should be avoided Warmth must be employed only as outlined above Vasodilating drugs are frequently not helpful and may be *harmful*

Surgical procedures should be considered for invading tumors and for aneurysms Within the first few hours after embolization surgery is indicated if a well trained team is available and the embolus is at the level of the *midfemoral artery or proximal to this* point

Surgical procedures are usually unnecessary in the treatment of early lesions of thromboangiitis obliterans and atherosclerosis Conservative management gives good results in most of these cases In advanced cases of gangrene amputation may be necessary This *should be regarded as a defeat of conservative* measures for any of numerous causes

After surgery the patient should be given the benefit of the same sound physiologic management used for medical cases The opposite *extremity which is frequently involved should* be given meticulous attention



C Five months later



D Heel shows clean granulation

The remainder of the foot was red swollen and cold

Pulses	Right	Left
Posterior tibial	0	0
Dorsalis pedis	0	0
Popliteal	0	0
Femoral	+	+
Oscillometric Readings		
Foot	0	0
Ankle	0 3	0 4
Calf	0 4	0 4
Thigh	1 2	1 0

The patient was instructed to stand by the side of her bed and take a few steps several times a day This was gradually increased and at the end of one week she walked to the bathroom with the help of two nurses An oscillating bed was set for a downswing of 15 degrees and no upswing above the horizontal The foot was covered with antibacterial ointment and wrapped in cotton It was dressed daily At the end of three weeks she was able to walk every hour for five minutes with help The temperature was defervescent Her narcotic addiction was reduced

She was sent home to a distant city to continue her treatment in collaboration with her personal physician An oscillating bed was

Case 2 Atherosclerosis Obliterans, Diabetes Mellitus, *Hypertension (170/100)*, Gangrene of Left Foot



A *Black gangrenous ulcers*



B One month later

Mrs R K (The New York Hospital # 705427) aged 69 presented the above complications Her diabetes had been treated for 18 years *Five months prior to admission* the left fifth toe had become infected It became gangrenous and was amputated The wound did not heal A spreading cellulitis developed The surrounding area became black Necrotic areas developed on the lateral side of the heel and on its posterior surface The patient was septic She was in great pain and had become addicted to narcotics Amputation of the leg had been advised by several consultants She had been in three different hospitals in an effort to save the leg

She was admitted to The New York Hospital on March 13 1955 She had not walked for the previous six months Examination showed extensive gangrene of the left foot Black circular areas were present at the heel tip (2 cm in diameter) lateral heel (3 cm) and lateral anterior surface (9 cm) and there was a large black slough over the stump of small toe amputation site as shown in Figure A



G Eleven months healing complete



H Section of femoral artery of similar patient

COMMENT This case illustrates the ability of the tissues to heal even with most advanced gangrenous lesions given sound therapy and adequate time It may be argued that this was too long a period for most patients and we agree but this patient desired to save the leg at all costs and it was possible to demonstrate that this could be accomplished

Figure H is another illustration of athero sclerosis This is a section of a femoral artery stained with hematoxylin eosin It is not taken from Case 2 but from a similar patient who came to autopsy

The internal elastic lamina is interrupted at many points A large portion of intima is replaced and covered by a hyalinized athero sclerotic plaque Some areas of this plaque show dense collagen material Other areas are more cellular and show a looser texture

The media is irregular in thickness Local necrosis has occurred in some areas where the smooth muscle has been replaced by col lagen

In the center of the section is a mass of eosinophilic material disintegrated red cells and platelets

Occasionally the disease progresses very rapidly in large vessels and occlusion occurs suddenly at a high level This is illustrated in Cases 3 and 4

Case 2 (Continued)



E Seven months



F Nine months

obtained She slept on it and remained on it during the day except when walking At two week intervals and later at one month intervals she returned to New York for follow up visits which included gradual debridement

One month later the large necrotic lesion over the anterior of the foot has localized as seen in Figure B The necrotic areas liquefied and slowly they were replaced by granulation tissue In Figure C taken five months after conservative treatment was started the black necrotic areas have sloughed and granulation has started The ulcer on the posterior tip of the heel has completely epithelialized Figure D is a close up of the lateral heel showing clean granulation Seven months after the first visit all necrotic areas were replaced by granulation tissue The fourth left toe was loose and without nerve innervation The fourth and fifth metatarsal bones had sloughed away in the necrotic debris Therefore this toe was removed during one visit This required no anesthesia and was practically painless (Fig E) Epithelialization took place (Fig F) Final healing was complete 11 months after the start of ambulant therapy (Fig G)

Case 3 Atherosclerosis Obliterans

He had great difficulty walking He reported to the emergency clinic and was sent to the vascular clinic by wheelchair

On examination he was *found to be acutely* ill In a horizontal position his legs and feet were white and cold as shown in Figure A A cyanotic rubor developed in both feet when they were in dependency (Fig B) No picture was taken with the legs in elevation because of danger to this patient who had such marked arterial insufficiency Pulses and oscillometric readings were as follows

Pulses	Right	Left
Posterior tibial	0	0
Dorsalis pedis	0	0
Popliteal	0	0
Femoral	0	0
Oscillometric Readings		
Foot	*0*	*0*
Ankle	0	0
Calf	0	0
Thigh	0 2	0 2
Pallor on elevation of feet	4+	4+
Rubor on dependency of feet	4+	4+

He was hospitalized at once He was placed on an oscillating bed *It was set so* that the feet did not rise above the horizontal position because of the marked pallor when the feet were elevated above this level The down swing for the feet was set at 15 degrees

He had smoked 40 to 60 cigarettes daily Tobacco was interdicted It required a supreme effort of will power to give up this addiction but he succeeded He has not resumed the use of tobacco *for the* past six years

Reflex heat, in the form of a heating pad was applied to the groin on alternate hours Alcoholic beverages were encouraged Sublingual nitroglycerin was given twice daily and later once daily

Ambulation was urged as soon as the decrease in pain would allow it

X ray studies were carried out A lateral view of the lower abdomen revealed a calcified plaque in the lower abdominal aorta The x ray study is shown in Figure C Note the calcification in the abdominal aorta

The results of injecting contrast media into the aorta are shown in Figure D This is an x ray AP view and it shows a block at the bifurcation of the aorta a narrowed channel in the right iliac and great development of collateral vessels Note the complete blockage of one iliac artery and narrowing of the other at the aortic bifurcation

A conservative regimen was decided upon His walking distance slowly increased until he was able to have bathroom privileges He returned to work This required eight hours a day on his feet He was instructed to protect himself against chilling In the winter he wears heavy underwear wool socks and stadium boots He has been cautioned *to avoid* heat to his feet

After six years he now walks two to three continuous miles slowly each day The collateral blood flow in his legs has increased enormously as evidenced by *the texture* and warmth of his skin His oscillometric readings are now

Oscillometric Readings	*Right*	Left
Foot	0 1	0 1
Ankle	0 4	0 8
Calf	0 6	0 8
Thigh	0 8	1 0

His generalized atherosclerosis obliterans and hypertension have progressed however *His right* ulnar artery *is not palpable* and the radial artery is weak His *left brachial* artery is partly occluded *However* the collateral flow is so good that he is symptom free in his arms Because of the occlusive disease blood pressure readings which had ranged as high as 220/110 can no longer be obtained

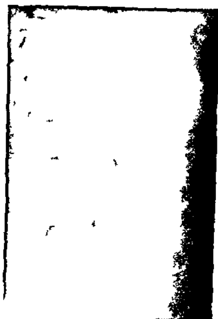
Case 3 Atherosclerosis Obliterans with Occlusion of Bifurcation of Aorta



A Legs show pallor in horizontal position



B Rubor of legs in dependency



C X ray of lateral abdomen



D X ray of abdomen AP view

Mr A P is a 58 year old watchman He has been employed at The New York Hospital for many years and had enjoyed good health up to the time of this episode except for a symptomless hypertension One cold winter's day while on duty outside the hospital entrance he suddenly experienced intense pain in his back His legs became numb and cold

Case 5 Atherosclerosis Obliterans with Gangrenous Ulcer, Healed by Conservative Therapy



Note black necrotic tissue



Two months later



One year later

Mr M W is a 65 year old automobile dealer who entered The New York Hospital (#669097) in *October* 1953 The diagnosis was atherosclerosis obliterans with occlusion of both popliteal arteries and a gangrenous ulcer on his shin which measured 7 by 3 cm It is well shown in the accompanying photographs He was a severe hypertensive (220/120) He did not have diabetes Vascular examination showed

	Right	Left
Pulses		
Posterior tibial	0	0
Dorsalis pedis	0	0
Popliteal	0	0
Femoral	2+	2+
Oscillometric Readings		
Foot	0	0
Ankle	0 3	0 2
Calf	0 4	0 6
Thigh	2 5	3 0
Pallor on elevation of feet	2+	2+
Rubor on dependency of feet	2+	2+

He had been fearful of walking for several months Ambulation was started at once The oscillating bed warm tub baths and reflex heat were employed He remained in the hospital for three weeks He continued his *treatment at home The health of the tissues* of his legs improved slowly The necrotic skin at the site of the lesion was dissected off

He was accustomed to crossing his feet when in bed The skin broke down over the outer malleolus at the point of pressure A second large necrotic ulcer formed Gradually both ulcers granulated He visited us at monthly intervals After six months all tissues appeared healthy but the two ulcers had not epithelialized They were covered with powdered blood cells A hard coagulum formed He was seen again in six months Epithelium had grown across and the skin was intact He continues to be well He walks one mile each day at a slow pace as a therapeutic measure

The upper left figure is a view of the lower leg Note the black necrotic tissue The upper right figure was taken two months later The necrotic tissue has sloughed A deep punched out ulcer is present which shows granulation The bottom figure was taken one year later The area is completely healed

Case 4 Calcification of Abdominal Aorta with Complete Obstruction

Mrs M is a housewife who was only 35 years of age at the time of the episode to be described

She had enjoyed excellent health She was a proficient athlete and a vigorous golfer Her menstrual pattern was normal in every respect

One day, while playing golf she started to walk up a hill Suddenly her legs would not hold her up She experienced sharp pain in her back and thighs She fell to the ground Subsequently she was removed to a hospital

On examination her legs were cold and pulseless Oscillometric readings and pulses were as follows

Pulses	Right	Left
Posterior tibial	0	0
Dorsalis pedis	0	0
Popliteal	0	0
Femoral	0	0
Oscillometric Readings		
Foot	0	0
Ankle	0	0
Calf	0	0
Thigh	0 1	0 2
Pallor on elevation of feet	4+	4+
Rubor on dependency of feet	4+	4+

She was placed in an oscillating bed with a 5 degree upswing and a 15 degree down swing Tobacco was interdicted an order which she obeyed with difficulty but successfully Reflex heat in the form of a heating pad to the abdomen was applied at alternate hours She was cautioned against applying heat to her feet Walking was commenced At first it was painful to even bear weight but soon she was able to take a few steps each hour

The illustration shows a radiograph of the abdomen in the lateral position Extreme calcification of the abdominal aorta is present opposite the bodies of lumbar vertebrae one two and three This undoubtedly was the site of a mural thrombus completely obstructing the aorta



Lateral radiograph showing extreme calcification of abdominal aorta opposite bodies of lumbar vertebrae one two and three

Studies of calcium and phosphorus metabolism were normal A complete endocrine study was normal Blood cholesterol was 195 mg

She left the hospital after two weeks At home she gradually increased her walking distance She is now able to walk any distance up to two miles at a moderate pace

The acute occlusion occurred in 1948 She has been followed carefully during the 11 intervening years The only illness that is worth noting is hyperthyroidism which developed in 1951 and which responded well to subtotal thyroidectomy

When vessels to segmental areas are occluded necrosis of defined areas may follow as in Case 5

Case 7 Atherosclerosis Obliterans

Case 7 Atherosclerosis Obliterans Diabetes Mellitus with Gangrene of Large Toe

Gangrenous toes



Separation of toe





Four months later following conservative therapy

Mr P McD The New York Hospital #783820 is a 58 year old municipal worker He presented himself with gangrenous toes as seen in the upper left figure His other leg had been amputated one year previously at another clinic

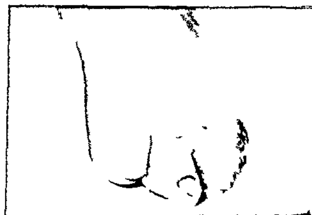
He was able to walk well with his artificial leg but had taken to bed when his toes pained him He had an occlusion of his femoral artery in Hunter s canal

He was treated as an ambulant patient and not admitted to hospital The diabetes was controlled by Orinase and diet Each hour walked for a minimum of five minutes gave up the use of tobacco Furacin solu dressing was applied daily to his toes necrotic toe separated at a line of dema tion as shown at the upper right The l was severed with scissors He was encour to walk and reported for weekly debriden Complete healing was obtained as show the bottom illustration taken four m later

Case 6 Atherosclerosis Obliterans, Diabetes Mellitus, with Gangrene of Large Toe and Foot



Gangrene of large toe and foot



After conservative therapy

Mrs G is a 68 year old Italian born housewife *She presented herself with a black gangrenous* toe and foot as seen on the left She had been confined to bed for five months

Examination showed occlusion of the popliteal artery Although the patient was brought in by ambulance weight bearing was started at once

She was not admitted to the hospital but was treated as an out patient She was instructed to stand up and walk every hour *Furacin soluble dressings were* applied daily The diabetes was controlled with insulin

She reported each week for debridement Walking was increased daily In a month's time she was able to carry on her normal activities The necrotic tissue was dissected free at the lines of demarcation

Healing was rapid and complete following conservative therapy



Radiograph of thigh showing calcification of the femoral and tributary arteries



Lateral view of knee showing calcification of popliteal artery



Lateral view of foot and ankle showing calcification of posterior and anterior tibial arteries

ARTERIOSCLEROSIS MÖNCKEBERG'S MEDIAL TYPE

Arteriosclerosis which involves the intima of arteries must be carefully distinguished from Monckeberg's sclerosis which concerns the medial layer of the vessels The latter *process does not reduce the caliber of the vessel* and is a benign process Case 8 illustrates this

Case 8 Arteriosclerosis (Monckeberg's Medial Type) with Extensive Calcification without Obstruction

This 77 year old man has extreme calcification of all large arteries They can be seen clearly outlined on the radiographs The top *figure shows calcification of the femoral* and tributary arteries The lower left illustration is a lateral view of the knee showing calcification of the popliteal artery The lower right figure is a lateral view of the foot and ankle and shows calcification of the posterior and anterior tibial arteries No major vessels are occluded *All pulses* are palpable Oscillometric readings are normal He had intermittent claudication in the calves after walking five blocks This was believed to be due to interference with the circulation in some minor branches supplying the calf muscles On conservative therapy he increased his walking distance to one mile

Case 9 Thromboangiitis Obliterans


Gangrene of large toe


Complete healing

Smoking was interdicted The oscillating bed was set at 0 degrees upswing 15 degrees downswing Fever therapy was given using intravenous typhoid vaccine every third to fifth day The foot was dressed daily The left figure was taken one month later when considerable improvement had occurred It was decided to ambulate this man and have him walk with crutches His intense pain was relieved After one month of this he was allowed to bear weight This was the first case of gangrene in our series in which weight bearing was tried He not only tolerated it well but improvement was more rapid He was discharged after two months hospitalization The toe had not yet healed but it was clean and granulating

He was seen thereafter at biweekly intervals Two months later healing was complete He returned to his job as a truck driver He has been seen twice yearly for the past seven years when as shown in the right figure complete healing has taken place His last examination was in January 1959 The foot was warm pink and dry The toe was intact He can walk any reasonable distance at a moderate pace without claudication

THROMBOANGIITIS OBLITERANS (BUERGER'S DISEASE)

This is an acute or subacute inflammatory disease involving the veins, arteries, and nerves. It occurs in young and early middle aged adults. Until recently it was largely confined to males but we are now seeing it more frequently in females. This is probably because of the increased and prolonged use of tobacco by women.

In our experience the disease occurs only in users of tobacco. It subsides when tobacco is interdicted. If smoking is resumed, even years later, it returns. It does not fit the usually accepted criteria for an allergy, nor is it entirely due to the vasoconstricting influence of nicotine. We feel that this disease is due to a peculiar constitutional idiosyncrasy to some substances in tobacco as yet not fully understood.

Frequently, episodes of phlebitis occur months or even years before arteries are occluded. In about 40 per cent of all patients phlebitis occurs at some time. The disease then goes on to involve digital arteries, most commonly in the toes. If the use of tobacco is not discontinued, large vessels such as femorals, iliacs, radials and ulnars eventually become involved. Finally, visceral vessels are often occluded, such as coronary and mesenteric arteries.

Case 9 Thromboangiitis Obliterans, Gangrene of Large Toe, with Healing

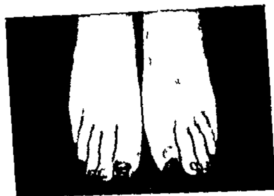
Mr. P. H., The New York Hospital #544287, admitted in July 1949, was a 38 year old truck driver. The diagnosis was thromboangiitis obliterans with gangrene of the right first toe and with occlusion of the right popliteal artery and of the right ulnar artery.

He had suffered from several episodes of superficial phlebitis during the prior two years. For three months the right foot had been increasingly numb and painful. A cramp consistently developed in the right calf after walking about 100 yards. There was constant burning pain in the right first toe which became cold and cyanotic. He had been hospitalized elsewhere. He had received procaine nerve blocks and intravenous procaine and the use of the oscillating bed. The lesion became progressively worse. He had been bedridden for two months.

On admission it was found that the right first toe was gangrenous (see the left figure). The foot was cold and pulseless. Pulses and *oscillometric readings* were as follows:

Pulses	Right	Left
Dorsalis pedis	0	2+
Posterior tibial	0	2+
Popliteal	0	2+
Femoral	Weak	2+
Radial	2+	2+
Ulnar	0	2+
Oscillometric Readings		
Foot	0	0.6
Ankle	0.3	3.5
Calf	0.6	3.5
Thigh	1.2	4.0

Case 10 Thromboangiitis Obliterans



Gangrenous ulcerations



Close up showing necrotic ulcerations



One year later



Section from another patient with similar disease

Case 10 Thromboangiitis Obliterans

Mr J S (The New York Hospital #662892) a 35 year old public school janitor was admitted to the hospital *in August 1953* The diagnosis was thromboangiitis obliterans with gangrene of toes and fingers The two top figures show the gangrenous ulcerations The digital vessels in these areas were thrombosed The top right figure shows a close up of the toes with necrotic ulcerations Seven months previously he had developed bilateral thrombophlebitis which had subsided after rest in bed for one month Two weeks prior to admission his toes became painful cold and blue The left index finger became swollen cold and ulcerated The right index finger soon became similarly cold and ulcerated These ulcerations had developed at the sites of minor abrasions

Examination showed partial blockage of the right radial artery and both ulnar arteries The dorsalis pedis and posterior tibial pulses were absent Oscillometric readings were zero bilaterally below the knees

Despite warnings the patient continued to smoke Both first toes developed gangrene A bilateral sympathectomy was done and as is the usual experience no improvement resulted until the patient gave up tobacco Walking was started Healing was then rapid The necrotic tissue sloughed on the toes and fingers He was seen at weekly intervals by Dr Ellen McDevitt in the clinic where his *wounds were dressed and gently debrided*

The patient returned to work Dressings were necessary for an additional six months before healing was complete He has been followed at frequent intervals in the clinic The lower left illustration was taken one year later Healing has followed the cessation of the use of tobacco together with conservative measures At present he has been followed for three and a half years since complete healing Oscillometric readings have increased markedly Hands and feet are warm pink and dry He works a full day performing his full duties as a janitor He walks long distances

The lower right figure is a microscopic section from another patient with a similar disease

The early lesions in this disease are thought to be an acute inflammation of all layers of the walls of veins and arteries Thrombi develop over the inflamed areas

This section of artery shows the internal elastic lamina embedded in a thrombus in various stages of organization Some areas are recent and *some fresh The dense black areas* are hemoglobin pigment Note the cellular reaction in the adventitia

Case 12 Thromboangiitis Obliterans Quadrilateral



Ulcers on right second and third fingers



Close up of ulcers

This 26 year old robust carpenter had signs of arterial blockage in all four extremities He was a heavy cigarette smoker The hands were red on dependency and pale on elevation The left photograph of both hands shows ulcers on the right second and third fingers The right photograph is a close up of the right hand and shows details of the ulcers The radial pulse is blocked on the right Oscillometric readings are greatly reduced in the wrist are zero in the hand and also are reduced in the left hand and wrist Dorsalis pedis pulses are absent

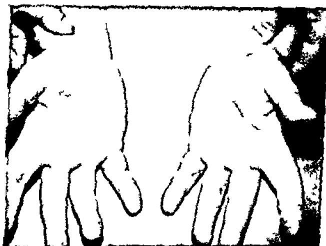
Case 11 Thromboangiitis Obliterans of Both Lower Extremities

Ulceration of right foot and large toe



Close up of ulcerated large toe and necrotic ulcer on dorsum of foot





Allen test

This 33 year old man had had an amputation of his left leg at age 19 from thromboangiitis obliterans He resumed smoking 14 years later The disease returned and he developed gangrenous ulcers of his right foot *and large toe as seen in the upper left figure* and changes in the vessels of the hands The upper right illustration is a close up of the *ulcerated large toe and necrotic ulcer on the* dorsum of the foot The bottom figure is an illustration of the Allen test The photograph shows the radial arteries blocked off by the *examiner's thumbs One hand is pink showing patency of the ulnar artery and palmar* arch The other hand is pale indicating a blockage of the ulnar artery

ARTERIAL EMBOLIZATION

Large clots form in the auricles or other chambers of the heart if the heart is fibrillating or if it is deformed because of mitral stenosis or another cause. Clots may also form over damaged areas of endocardium as in myocardial infarction. These clots may break off and flow out into the systemic circulation lodging in the brain, viscera or extremities.

When a large clot occludes the bifurcation of the abdominal aorta it is known as a "saddle embolus." Such a case is illustrated in the following history.

Case 14 Embolus to Lower Extremities



Necrotic black toe and dependent rubor



Toe separated during a dressing



Complete healing

Mrs M S The New York Hospital #696317 had an embolus of the lower abdominal aorta and gangrene of the foot. This 51 year old factory worker had been well until six weeks before her admission in November 1954. At that time she developed severe angina pectoris. This gradually increased. It culminated in an episode of severe substernal pressure which led to admission in another hospital. A diagnosis of myocardial infarction

Case 13 Thromboangiitis Obliterans in a Woman



Dependent rubor of affected foot



*Granulating stump of num*ber four toe

This 39 year old housewife smoked 60 cigarettes daily. Four years ago she bruised her left foot and an ulcer formed on her left fourth toe. The ulcer would not heal and the *toe was amputated and a sympathectomy was* performed. The stump would not heal. She had not been told to stop smoking. This interdiction was enforced and healing followed.

The left figure shows intense dependent rubor *of the affected foot compared to the normal* color of the other foot. The right figure is a close up of the toes showing granulating *stump of fourth toe. Pulses are absent in the* foot. Oscillometric readings are reduced up the leg.

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was established Anticoagulants were not given On the fourteenth day while straining on a bedpan she developed a sudden pain in her left leg The leg rapidly became cold and blue She was then transferred to The New York Hospital

Examination showed cold pulseless cyanotic legs Femoral pulses were absent Our diagnosis was embolization of the lower abdominal aorta from a mural thrombus secondary to myocardial infarction Treatment consisted of anticoagulant therapy with the use of an oscillating bed and reflex heat The legs became warmer but the left large toe became black and mummified Collateral flow developed down into the foot All areas became pink and warm except for the black toe The patient was ambulated and discharged She came to the clinic weekly where she was treated by Dr Ellen McDevitt In March (four months after her embolic phenomenon) the necrotic toe was gently twisted off with a thumb forceps The stump quickly epithelialized A follow up for three and a half years shows continuous improvement in collateral circulation She works at her factory job as before

The accompanying photographs show the necrotic black toe and the dependent rubor The patient was encouraged to walk long distances slowly Arterial flow continued to improve She was treated as an out patient at weekly intervals Perfect healing was obtained

If such a patient is seen within a few hours after embolism blocking the aorta iliac or femoral arteries of the lower extremities or the subclavian axillary arteries serious consideration should be given to immediate embolectomy The general condition of the patient must be considered before such an operation A recent myocardial infarction is usually considered as a grave surgical risk

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2

Venous Diseases

1 VENOUS INSUFFICIENCY

Patients with venous insufficiency like most patients with diabetes cannot be cured but they can be maintained in good health provided they follow certain hygienic measures

In the upright posture a column of blood extending from the right auricle to the ankle may exert as much as 100 mm of mercury of hydrostatic pressure If this enormous head of pressure were transmitted directly to surface veins they would rupture Counteracting this are valves inside the veins and the turgor of muscles and tissues outside

Some persons are born with weak venous valves Rarely they may be absent in certain venous segments such as the communicating veins between the deep and the superficial systems of the legs In others the vein walls are weak and after years of pressure they sag outward at the sites of the valves preventing the leaflets from closing tightly and thus making them incompetent These are the key factors in the development of varicose veins which occur frequently in some families After years of standing the valves become defective under the strain The hydrostatic pressure is then passed along distally to the next valve In time it too may give way When the pressure is communicated to a surface vein it becomes tortuous or varicose upon standing Reversal of direction of blood flow may occur in such a vein The enules which it drains are then subjected to increased pressure The usual drainage of fluid from the tissues is interfered with Waste products accumulate in the tissues especially about the ankles Red blood cells leak out into the tissue spaces and disintegrate They leave behind their iron pigment which stains the tissues a rust color Tissue cells become atrophic microorganisms thrive and a condition known as "stasis dermatitis" occurs Fluid accumulation results in intracellular edema Finally the skin dies and ulceration follows

The ideal treatment would be to replace the defective valves This is a project for the future At present all attempts to do so have failed owing to the ease of thrombus formation at the site of the prosthesis

Elevation of the leg reduces the venous pressure to zero or below and results in rapid venous drainage This utilization of the aid of gravity can usually be accomplished for one third of each 24 hours by having the patient sleep with the feet elevated six inches by placing blocks under the legs of the bed This is more comfortable and more effective than elevating the legs on a pillow or raising the *mattress*

The increased venous pressure on the surface of the legs can be counteracted by the *compression* of an elastic support Ideally this support should just balance the increased pressure Empirically it has been found that

this may usually be accomplished by exerting 50 mm of mercury pressure at the foot and ankle gradually diminishing to a pressure of 20 mm just below the knee To do this many measurements are needed The circumference should be measured at one inch intervals from toes to knee

Stockings that come above the knee often bind the popliteal space and give a tourniquet action This occurs especially when the knee is bent Knee length stockings are usually preferable The severe effects of venous insufficiency manifest themselves in the lower half of the leg It is in this area that compression is needed Caution must be used in the use of compression when there is evidence that the patient is also suffering from impaired arterial circulation Here the physician must use good judgment weighing carefully the relative seriousness of the two conditions

Elastic girdles and round garters impede venous flow and must not be worn in this condition

Walking in water or swimming exerts a beneficial effect on the venous flow The water pressure is maximum at the feet and less and less up the body—ideal for clearing the tissues of stagnant lymph Patients suffering from venous insufficiency almost uniformly report great improvement after a summer of swimming

Postphlebitic Syndrome During the acute phase of thrombophlebitis the valves of the involved veins often become enmeshed in the inflammatory action and thus are deformed or destroyed Later after varying periods of time the absence of these valves manifests itself in the involved leg by the signs of chronic venous insufficiency This damage is reduced if the patient uses an elastic stocking or support from the first day of ambulation

Walking with a well fitted elastic stocking in place is beneficial to the leg with venous insufficiency The muscular contraction gives a pumping action to the deep veins and supplants the dynamic action of valves Standing still however is a great strain on the venous flow Sitting with legs dependent for long periods of time is to be avoided We instruct patients to walk about every half hour when on long plane train or automobile trips This advice also pertains to long meetings card games or other periods of prolonged sitting

For months and even years after an attack of phlebitis the legs are subject to episodes of pains and aches These are most apt to occur *during a fall of barometric pressure* and enable the patient to forecast a period of bad weather Patients are apt to worry and fear that there has been a return of acute phlebitis We instruct them to elevate their legs markedly and apply hot moist packs Such pain often disappears very quickly and should be relieved *in a period of at most two hours* If it lasts longer than this a recurrence of phlebitis should be considered and a physician should see the patient

Sclerosing by the Empty Vein Technic Sclerosing injections have been employed for many years by the profession to obliterate defective superficial veins Various substances have been used They are usually effective for a few months but recanalization takes place and the varix sooner or later returns Local pigmentation frequently occurs at the site of the injection

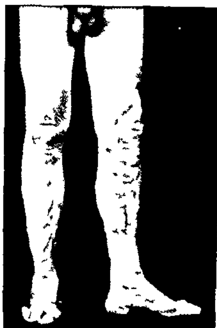
We use a technic of injecting varices *after they have been emptied of blood* In this manner usually no thrombus is formed The intima of the vein becomes inflamed The vein contracts immediately after injection The walls of the vein grow together and the vein becomes a fibrous cord If proper elastic stockings are worn thereafter there is less tendency for varices to recur

SURGICAL MEASURES

It is beyond the scope of this book to enter into a dissertation of the various surgical measures employed In a general way it may be stated that the aim of surgery is to eliminate the incompetent veins that penetrate the deep fascia and connect the deep venous channels with the superficial veins (7) The main veins that do this are 1 the great saphenous vein which extends from the groin down the inner side of the leg to the ankle 2 the short saphenous vein which extends from the popliteal space down the outer side of the leg 3 several short veins in Hunter s canal 4 communicating veins just above the inner and outer malleolus 5 at times other communicating veins along the foreleg

When a superficial vein is connected to a deep vein via a communicating vein that is incompetent it can often be demonstrated that the blood flows in the opposite direction when the patient is in an upright position Careful ligation of these incompetent veins flush to their point of origin from the deep vein is the fundamental aim of surgery The eradication of dilated varices by such measures as "stripping" is usually done as well It is our experience that patients who wear well fitted elastic stockings after surgery get the best results

Case 15 Varicose Veins



Dilated varicose veins



One year follow up

Mr W G (The New York Hospital #536425) is an 80 year old executive He noted varicose veins about 20 years ago These gradually increased in size until they became markedly enlarged throughout the long and short saphenous systems as shown in the left figure

A fitted elastic stocking was prescribed with graded pressure A series of three injections of Sylnasol was given at monthly intervals using the empty vein technic The right figure was taken one year later Note the disappearance of the enlarged veins He must continue to wear his gradient elastic stocking to avoid recurrence

Case 16 Varicose Veins, Stasis Dermatitis, Ulceration



Before treatment



Six month follow up

Mrs E K is a 50 year old housekeeper Prominent varicose veins developed at age 25 They gradually increased in size Pigmentation formed at the ankle together with a plexus of veins The skin of the left inner malleolus became rough and itchy About two years ago a painful ulcer formed

The left photograph shows the prominent varices pooling pigmentation stasis eczema and ulcer formation of two years duration The flow of blood in these dilated veins was retrograde in changing the posture of the patient from the reclining to the standing position

Fitted elastic stockings with graded pressure were prescribed The defective veins were obliterated, using the empty vein technic In particular one communicating vein draining the ulcer was injected The right figure was taken six months later Note the disappearance of the dilated veins the healing of the ulcer and the return of skin color

Case 17 Chronic Venous Insufficiency, Varicose Veins, Ulceration



Note venous plexus ulceration pigmentation and devitalized skin



Two month follow up

Mr O H (The New York Hospital #788253) is a 60 year old butcher For some 10 years he has had varicose veins probably a complication of standing still for many hours each day in his shop

Five years ago ligations of the long and short saphenous systems improved his legs temporarily He did not wear elastic stockings Pigmentation plexus formation stasis dermatitis and finally painful ulceration developed

He was hospitalized and skin grafted The ulcer recurred His surgeon referred him to us and the left photograph was taken Note the venous plexus ulceration pigmentation and devitalized skin

A communicating vein was palpated slightly above the ulcer It was sclerosed using the empty vein technic He was fitted with a pressure gradient elastic stocking Healing was rapid as shown in the right photograph He returned to his work as a butcher and has remained well

Case 18 Chronic Venous Insufficiency Varicose Veins, Ulceration

Ulceration and devitalized skin



Close up of ulcerated area





Six month follow up

Mr W C (The New York Hospital #773617) is a 56 year old metal worker Ten years ago he suffered fractured tibias During his enforced immobilization in a plaster cast he developed thrombophlebitis This is a common complication of fractures

The phlebitis destroyed the deep venous valves and left him with chronic venous insufficiency Pigmentation and devitalization of the skin soon followed together with varicose veins A slight bump of his ankle led to ulcer formation

The upper left figure shows the ulceration and devitalized skin The upper right figure is a close up of the ulcerated area Two defective veins with retrograde flow led to the ulcer They were obliterated by the empty vein technic He was fitted with a graded pressure elastic stocking The lower figure taken six months later shows excellent healing and return to health of the devitalized skin

2 THROMBOPHLEBITIS

Inflammation in or around a vein with clot formation is known as thrombophlebitis When intravascular thrombus formation is due to venous stasis or to alterations in the blood which increase the clotting tendency and is only followed secondarily by inflammation it is frequently termed phlebothrombosis It is often difficult to classify a case as belonging clearly to either group While in some patients the process at the time of onset is of one or the other type after several days inflammation and thrombosis are usually coexistent

Classification and Etiology The classification presented here is from the Nomenclature of Diseases of the Blood and Lymph Vessels prepared by a committee of the New York Heart Association (1)

I Thrombophlebitis and venous thrombosis (phlebothrombosis)
- (a) Primary
  - (1) Thromboangiitis obliterans
  - (2) Recurrent or migrating (without arterial lesions)
  - (3) Essential
- (b) Secondary to
  - (1) Mechanical injury (contusion laceration surgery)
  - (2) Muscular effort or strain
  - (3) Chemical injury (sclerosing agents drugs solutions for diagnosis)
  - (4) Inflammatory or suppurative lesions—infectious diseases
    - a Tuberculosis syphilis actinomycosis
    - b Other bacteria (to be specified)
  - (5) Infectious diseases
  - (6) Severe ischemia
  - (7) Chronic disease of vein wall (varices phlebosclerosis) (Later complications—varicose or postphlebitic ulcers)
  - (8) Blood dyscrasias (polycythemia vera leukemia pernicious anemia)
  - (9) Congestive heart failure
  - (10) Carcinoma

2 Neoplastic invasion of vein

3 Venous compression—with or without thrombosis of thrombophlebitis due to
- (a) Gravid uterus
- (b) Neoplasm
- (c) Aneurysm
- (d) Scar tissue
- (e) Scalenus syndrome
- (f) Fractures and dislocations
- (g) Increased intraabdominal pressure (ascites etc)
- (h) Extrinsic pressure (tight girdles circular garters poorly made trusses etc)

The wide variations in etiology and mechanisms are clear in the above classification The underlying factors in any one case may be further clarified by study Trauma or damage to a vein wall may initiate a thrombus formation especially when a substance to be injected is irritating in nature such as arsenicals ascorbic acid glucose aureomycin and certain of the opaque media used for x ray visualization of the vascular tree In addition it is now established that cortisone ACTH and certain oral arsenical compounds should be used with great caution in the presence of any condition which exhibits a thrombotic tendency It is recommended that crystalline trypsin not be used in clinical practice since it exerts a thrombosing action prior to the anticoagulant action The anti inflammatory action which is claimed for it is inadequately documented at present Stasis *per se* may be the inciting factor Increased tendency to clot formation in itself may lead to thrombosis as in cases of pancreatic neoplasm

Diagnosis The finding of a red tender hot painful cord in the course of a vein is the pathognomonic sign Frequently however a deep vein is involved without superficial signs In such cases reliance is placed on indirect findings to make a diagnosis These are dependent edema and cyanosis of the affected limb the dilatation of collateral veins deep

muscle tenderness pain on passive motion or stretching fever and constitutional symptoms Often edema and swelling may be masked and revealed only by accurate limb measurements (circumference) comparing equal levels in opposite extremities

TREATMENT

1 Elevation An involved extremity should be elevated approximately four inches above the level of the heart This is especially indicated and may be increased to eight inches in the presence of definite edema The purpose is to facilitate drainage of static lymph and venous fluid from the extremity This measure adds greatly to the patient's comfort Elevation is best accomplished by raising the foot of the bed six to eight inches on blocks The use of pillows alone too frequently results in elevation of the knee above the foot and therefore interferes with proper drainage distal to the knee In the presence of occlusive arterial disease it is unwise to elevate the extremity In such a paradoxical situation the use of an oscillating bed will help to reduce the edema safely Otherwise the extremity should be kept in a horizontal position

2 Rest Although it is important to keep the patient active as a preventive postoperative measure in order to discourage the production of clots and the development of thrombophlebitis once thrombophlebitis has been recognized the patient should be placed at bed rest until the signs of active phlebitis have subsided

3 Heat Gentle warmth is one of the best means of releasing venospasm and producing analgesia Arteriospasm is also a frequent complication of phlebitis A warm moist pack gives relaxation of both arterial and venous trees comparable to that produced by lumbar ganglionic block It may be maintained over long periods and is a great deal simpler The packs are at first applied for 20 out of each 24 hours decreasing the time as the condition improves It is wise to cover the skin with petroleum jelly or cold cream to prevent maceration from the long exposure to moisture

4 Nerve Blocks Rarely it may be necessary to perform sympathetic nerve blocks to release vasospasm It is considered by many to be hazardous to perform this procedure while active anticoagulant therapy is in progress The decision must therefore be made early

5 Analgesia and Treatment of Cramps Night cramps may frequently be relieved by 0 2 gm of quinine sulfate at bedtime or by 0 05 gm of benadryl

6 Antifungus Therapy Dermatophytosis of the toes and feet because it is a portal of entry for microorganisms should be actively combated by the usual means ($KMnO_4$ foot soaks 1/10 000 Whitfield's ointment half strength or undecylenic acid ointments) Over treatment should be avoided

7 Constitutional Treatment Dehydration must be avoided particularly since it increases the tendency to thrombus formation Unless there is some cause of sodium retention e g renal or cardiac disease edema will not increase from a liberal fluid intake

8 Anticoagulant Therapy The use of anticoagulants requires well trained teams of workers and laboratories prepared to provide scrupulous service Unless the laboratory's technic for determination of the prothrombin time is properly standardized and can be depended on use of coumarin or phenylindanedione derivatives should not be attempted When home therapy is undertaken provision should be made for frequent determination of the prothrombin level by reliable laboratory technicians

Barker and others (2) at the Mayo Clinic clearly demonstrated the great value of dicumarol in the prevention of pulmonary emboli in treating cases of phlebitis Zilliacus (3) and others have shown a similar beneficial effect with heparin These results have been reproduced in many clinics throughout the world

2 THROMBOPHLEBITIS

Inflammation in or around a vein with clot formation is known as thrombophlebitis When intravascular thrombus formation is due to venous stasis or to alterations in the blood which increase the clotting tendency and is only followed secondarily by inflammation it is frequently termed phlebothrombosis It is often difficult to classify a case as belonging clearly to either group While in some patients the process at the time of onset is of one or the other type after several days inflammation and thrombosis are usually coexistent

Classification and Etiology The classification presented here is from the Nomenclature of Diseases of the Blood and Lymph Vessels" prepared by a committee of the New York Heart Association (1)

1 Thrombophlebitis and venous thrombosis (phlebothrombosis)
   (a) Primary
      (1) Thromboangiitis obliterans
      (2) Recurrent or migrating (without arterial lesions)
      (3) Essential
   (b) Secondary to
      (1) Mechanical injury (contusion laceration surgery)
      (2) Muscular effort or strain
      (3) Chemical injury (sclerosing agents drugs solutions for diagnosis)
      (4) Inflammatory or suppurative lesions—infectious diseases
         a Tuberculosis syphilis actinomycosis
         b Other bacteria (to be specified)
      (5) Infectious diseases
      (6) Severe ischemia
      (7) Chronic disease of vein wall (varices phlebosclerosis) (Later complications—varicose or postphlebitic ulcers)
      (8) Blood dyscrasias (polycythemia vera leukemia pernicious anemia)
      (9) Congestive heart failure
      (10) Carcinoma
2 Neoplastic invasion of vein
3 Venous compression—with or without thrombosis of thrombophlebitis due to
   (a) Gravid uterus
   (b) Neoplasm
   (c) Aneurysm
   (d) Scar tissue
   (e) Scalenus syndrome
   (f) Fractures and dislocations
   (g) Increased intraabdominal pressure (ascites etc)
   (h) Extrinsic pressure (tight girdles circular garters poorly made trusses etc)

The wide variations in etiology and mechanisms are clear in the above classification The underlying factors in any one case may be further clarified by study Trauma or damage to a vein wall may initiate a thrombus formation especially when a substance to be injected is irritating in nature such as arsenicals ascorbic acid glucose aureomycin and certain of the opaque media used for x ray visualization of the vascular tree In addition it is now established that cortisone ACTH and certain oral arsenical compounds should be used with great caution in the presence of any condition which exhibits a thrombotic tendency It is recommended that crystalline trypsin not be used in clinical practice since it exerts a thrombosing action prior to the anticoagulant action The anti inflammatory action which is claimed for it is inadequately documented at present Stasis *per se* may be the inciting factor Increased tendency to clot formation in itself may lead to thrombosis as in cases of pancreatic neoplasm

Diagnosis The finding of a red tender hot painful cord in the course of a vein is the pathognomonic sign Frequently however a deep vein is involved without superficial signs In such cases reliance is placed on indirect findings to make a diagnosis These are dependent edema and cyanosis of the affected limb the dilatation of collateral veins deep

with the foot of the bed elevated on blocks six to eight inches high

2 Compression When up and about he should wear a firm well fitted elastic stocking extending from just above the toes to one inch below the knee These are provided for him when he begins ambulation

3 Hydrotherapy Swimming or walking in deep water should be encouraged

In other words every effort should be made to prevent the edema from becoming static or fixed until the compensating venous circulation is able to develop

The experimental work of Burt and his colleagues (4) indicates that clots are absorbed *more quickly if the patient receives anticoagulant therapy*

TECHNIC OF ADMINISTRATION OF ANTICOAGULANTS

1 The prothrombin time is *determined* by the Quick or Link Shapiro technic before the first dose is given *Normal readings are* 12 *to* 13 seconds by the Quick method and 13 to 17 seconds by the Link Shapiro method

2 If the prothrombin time is normal or lower 300 mg of dicumarol is administered orally in one dose If emboli have occurred heparin may be started immediately and continued until the dicumarol effect is manifested

3 Each day the prothrombin time is determined and reported to the physician in charge of the patient before the dicumarol dosage for that day is decided on

4 The second day 150 to 200 mg of dicumarol is given and thereafter it is given in doses averaging 25 to 100 mg daily (depending on the patient s sensitivity to the drug) An effort should be made to keep the prothrombin time one and one half to twice the normal time

5 If the *prothrombin time reaches* 35 seconds dicumarol is discontinued until the prothrombin time drops below 30 seconds after which it is given again cautiously *in* daily doses of 25 to 100 mg (These prothrombin time values are based on studies using the Link Shapiro method) The time may increase for several days after dicumarol is discontinued and then return toward normal

6 If the prothrombin time reaches 60 or 70 seconds hemorrhagic manifestations may occur and one must be alert to this possibility At a level of 60 to 65 seconds 10 mg of vitamin K_1 should be given orally Minor hemorrhagic *manifestations* such as purpuric spots minor oozing from the gums and some red blood cells in the urine are occasionally seen They are considered to be signals for alert but not necessarily an indication to discontinue the anticoagulant therapy More severe hemorrhagic manifestations can usually be checked by giving vitamin K_1 orally The objective is to keep the prothrombin level between 25 and 35 seconds especially during the first two or *three weeks* The dosage is *then tapered off slowly* over a period of from one to two weeks permitting a gradual return of the prothrombin time to normal Dicumarol has been continued in most of our cases until the patient has resumed his accustomed activities This is usually from 15 to 30 days after the last episode of thrombosis or embolism

When embolic phenomena have already occurred it is usually advisable to give heparin during the first 24 to 48 hours until *the effect of the first dose of dicumarol* is established The subcutaneous injection of concentrated heparin (10 000 u per ml) is very satisfactory With an average dose of 15 000 u adequate anticoagulant activity is obtained for 8 to 12 hours

Sublingual administration of heparin has failed completely to produce any change in the clotting time in tests run in our laboratory

AFTER CARE

During the acute phase of thrombophlebitis the valves of the *involved veins often become enmeshed in the inflammatory action* and are deformed or destroyed Months or years later the absence of these valves shows itself in the involved leg by the *signs of* chronic venous insufficiency They are varicose vein formation congestion of dependent tissues especially at the ankle hemosiderin deposit plexus formation stasis dermatitis and ulceration of the skin

To prevent this unfortunate chain of events the following instructions are given to the patient when he leaves the hospital or assumes his normal mode of living

1 Elevation The patient should sleep

Case 19 Thrombophlebitis

On examination the long saphenous vein was found to be thrombosed from mid thigh to calf Large clots filled the dilated varices Some measured as much as one inch in diameter The foot and ankle were swollen with 3+ pitting edema The calf at its maximum circumference measured two inches greater than the other calf On dependency a deep cyanosis developed When standing collateral veins became prominent over the hip Weight bearing produced pain in the foot and leg Passive flexion of the foot elicited calf pain

She was placed at bed rest The foot of the bed was elevated on six inch blocks Hot moist packs were applied Anticoagulants were administered 15 000 u of heparin was given subcutaneously every 12 hours The concentrated form was used (10 000 u per ml) On the third day dicumarol was started Therapeutic levels of prothrombin time were reached on the fifth day Heparin was discontinued and dicumarol was maintained at a daily dose that kept the prothrombin time between 25 and 35 seconds

Pain and tenderness subsided rapidly but the hard clots persisted for many weeks On the tenth day her temperature returned to normal She was ambulated A well fitted elastic stocking was made It extended from the toes to one inch below the knee On the twentieth day she was discharged from the hospital

She was instructed to 1 walk about with the stocking on 2, sleep with the foot of the bed elevated 3 swim or walk in deep water as often as possible 4 elevate the feet on a footstool when sitting 5 avoid tight garments and sitting for long periods of time as in a train or plane trip Dicumarol was continued for an additional six weeks Each week her prothrombin time was checked and she was cautioned to expect aches and pains in her legs from time to time especially when there was a sudden fall in barometric pressure After six weeks the dose of dicumarol was gradually decreased then stopped in two weeks

She returned to her position as a clerk One year later her leg showed the varicose veins as before but no additional signs of venous insufficiency had developed as her elastic stocking protected her She was told that her venous insufficiency could be improved only by surgery but that if she followed the regimen we outlined she might escape further difficulty Her elastic stocking had to be replaced every three months

The illustration shows large areas of phlebitis in the internal saphenous vein

Case 19 Acute Superficial Thrombophlebitis in Varicose Veins with Extension to Deep Veins



This patient is a stout middle aged white woman Varicose veins are prevalent in both sides of her family She inherited weak venous valves *After years of standing these valves* ruptured one after another until the entire venous system from heart to ankle was devoid of this protection Under this increased head of pressure the superficial veins became widely dilated and varicose

One day she took a long automobile trip She wore a girdle In the sitting position this girdle bunched up in the groin and acted as a tourniquet to impede blood flow The stagnant *blood clotted in the veins*

The following day she noticed red tender hot cords along her inner *thigh They quickly* extended down to the knee and calf The following day *the entire leg was swollen* due to blockage of the deep veins She developed a fever

3 PULMONARY EMBOLISM AND INFARCTION

Pulmonary embolism is very frequently overlooked or misdiagnosed in clinical practice Surgeons are apt to confuse it with postoperative atelectasis or pneumonia (Case 23) until active thrombophlebitis becomes evident General practitioners are prone to confuse it with virus pneumonia until the patient calls attention to the pain in his leg We have treated two physicians each of whom had been so diagnosed by their confreres One had four attacks" before the phlebitis in his saphenous vein was given its proper significance Patients with rheumatic heart disease have been treated for multiple episodes of pneumonia over a period of years until embolization finally has been correctly diagnosed as in the instance of Case 24 On careful study patients diagnosed as having pulmonary disease have occasionally proved to have had a myocardial infarction with pulmonary embolism from a mural thrombus

Farmer and Smithwick (1) have reported on 7 343 consecutive surgical admissions In 95 of these pulmonary infarction was recognized in 25 pulmonary infarction was the first indication that phlebitis was present

The terms pulmonary embolism and pulmonary infarction are not synonymous A pulmonary embolism means that a foreign body most commonly a portion of a thrombus has lodged in a branch of a pulmonary artery with or without changes in the lung tissues supplied by that branch If there is a profound change in the lung tissue which progresses to necrosis the term infarction is used Emboli in the absence of infarction do not usually produce detectable x ray changes unless opaque visualization studies are performed If the disturbed area is near the lung surface however changes in physical signs will be noted especially the finding of rales or a pleural friction rub The work of Kjellberg (5) aids in understanding this phenomenon He subjected a series of dogs to artificial pulmonary embolization No roentgenographic changes in the lungs were observed in uncomplicated pulmonary embolism and no infarction occurred as long as the circulation through the bronchial artery was intact

The recent work of Knisely (17) and his group suggests that mechanical blockage is the cause of death in pulmonary embolism rather than reflex vasospasm Infarction usually extends to the surface of the lung The irritated pleura reacts by an outpouring of fluid Pleural effusion is thus a frequent finding Blood exudes through damaged capillary walls into alveolar spaces Therefore hemoptysis is also a common finding

Symptoms *Pain* is often the first and most compelling symptom Two types of pain are encountered 1 that produced by reaction of the pleura overlying the infarction and 2 severe anterior chest pain often indistinguishable from that of myocardia It sometimes produces "splinting" and difficulty in breathing Some workers believe that this pain is the result of an actual diminution in the coronary flow secondary to the profound disturbances in the local hemodynamics (2 3 4) Others hold that this pain is located in the pulmonary artery itself This question has not been settled as yet

Dyspnea is common It is due to the splinting caused by pain rather than by the dysfunction of the rather small amount of lung tissue usually involved With a large embolus death may ensue instantly or within a very few minutes

Cough with hemoptysis of slight or marked degree is a most important corroborative sign Without it a diagnosis is often difficult to establish We have seen patients how

Case 20 Chronic Recurrent Low Grade Phlebitis



Chronic phlebitis



Six month follow up

Miss G is a 55 year old clerk About 20 years ago she developed a superficial phlebitis in her left leg following slight trauma It had recurred several times Each recurrence required bed rest for as long as two months The long saphenous vein was ligated Varices were interrupted and removed on several occasions The episodes of phlebitis recurred

The left figure shows her on her admission to The New York Hospital (#755214) Hard indurated tender lumps were present from the ankle to the mid calf Large superficial veins were found draining into this area The blood flow was reversed in these veins They were sclerosed using the empty vein technic The foot of the bed was elevated Heparin was given for two weeks A well fitted elastic stocking was made

She returned to work greatly improved but with some induration still present She continued to treat it at home applying hot moist packs to the lesion for one hour each night It took six months to clear the remaining inflammatory areas She must continue to wear her elastic stocking the rest of her life and follow the regimen outlined for Case 19 Her prognosis for continued activity is excellent

In the left figure note the brawny induration above the ankle This is the site of a low grade chronic phlebitis The dark areas above this are sites of hemosiderin deposits and dilated varices Stasis eczema and ulceration are present

The right figure taken six months later shows the return of the tissues to health

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ever with convincing evidence of pulmonary *infarction without hemoptysis* and even in the absence of positive x ray findings

The emotional reaction may be one of panic and fear of disaster This depends on the patients personality but the panic reaction is sufficiently common to warrant comment

Signs *Fever* will depend on the degree of infarction or secondary pneumonitis and the presence or absence of phlebitis at the site of origin Some fever is usually present and it may be marked

Heart Rate and Rhythm Tachycardia is present in proportion to the fever Arrhythmias are often present and are responsible for many emboli especially in rheumatic heart disease Any patient who is fibrillating is a candidate for pulmonary emboli We have seen a patient in normal rhythm have an episode of paroxysmal fibrillation discharge an embolus and revert to normal rhythm in a period of several hours Emboli may be discharged from hearts in normal rhythm especially if there is evidence of valvular damage It is our impression that it is the recently formed thrombus or recently propagated new clot superimposed upon old clot in a heart or vein that tends to break off and embolize

Signs of consolidation dullness to percussion bronchial breathing rales and a pleural friction rub may be present if the infarct is large enough Over areas of pleural effusion diminished breath and voice sounds will be heard As the effusion increases flatness develops Careful examination should at least show rales somewhere in the chest to support the diagnosis It is however ill advised to ask the patient to breathe deeply for this purpose This creates a negative pressure and may suck another embolus loose from the venous system

Findings *Laboratory* Leukocytosis with an increase in polymorphonuclear leukocytes is usually present depending on the presence or absence of phlebitis and pneumonitis The sedimentation rate increases rapidly during the first week

X ray The presence of a shadow in the lung fields *will be found only if some degree* of infarction has occurred The shape size and density of the shadow depend on the angle which the infarct makes to the direction of the x rays Thus shadows vary from small hazy mottled areas to large dense segments If seen in several different areas during the period of observation the diagnosis is further substantiated The classic pie shaped area *while sometimes seen is not found in the* majority of cases Some degree of pleural effusion is usually seen in the x ray films some time during the course of the illness

Electrocardiographic A large pulmonary embolus by blocking a generous percentage of pulmonary arterial flow produces an increase in pulmonary artery pressure This in turn is reflected in the heart by dilation of the right ventricle and what is termed "right heart strain" The electrical changes are a shift of the axis to the right with clockwise rotation This gives the following changes in the leads

I Deep S depressed ST
III—Q elevated ST
aVR—R elevated ST
aVL—depressed ST inverted T
aVF—elevated ST
V and V—R prominent elevated St VAT prolonged
V V V—ST depressed S marked in V V

Variations from this pattern depend on the *size and location of the embolus*

Complications *Cor pulmonale* Acute dilation of the right ventricle and pulmonary conus may follow massive pulmonary emboli These changes may produce signs which can be recognized on physical examination Recurrent pulmonary emboli may produce chronic cor pulmonale Distention and increased pulsations of the veins of the neck are common signs Increase in pulsation may also be noted in the second and third inter

spaces to the left of the sternum The second pulmonary sound may be heard over the area of increased pulsation Gallop rhythm heard best to the left of the sternum may be present

Shock "Peripheral circulatory collapse" may be present and require emergency measures While we recognize that shock is a variable manifestation most patients we have encountered in shock from pulmonary embolization display cold moist pale hands moist face cyanosis weak feeble rapid pulse and hypotension

Differential Diagnosis Space does not permit discussion of all conditions which must be considered in a differential diagnosis The condition will be recognized more often if the possibility of pulmonary embolus is kept in mind All patients with cardiac arrhythmias rheumatic heart disease recent myocardial infarcts recent surgery or fractures pregnancy polycythemia and phlebitis are apt to have pulmonary emboli

PREVENTION AND TREATMENT

In a sense pulmonary embolism may be likened to lightning It strikes most often with great suddenness and if the patient survives the initial shock he has a good chance of surviving that particular embolus It is the emboli yet to be delivered that are most to be feared indeed many large fatal pulmonary emboli are preceded by small ones As Barker (6) pointed out if all fatal pulmonary emboli preceded by minor emboli could be prevented the death rate would be strikingly reduced Autopsy statistics at The New York Hospital showed a rate of 10 per cent pulmonary embolism in the years before anticoagulants were used

For this particular syndrome the best treatment is definitely prevention A review of a large number of case histories has shown that a greater awareness or a more careful study on the part of the physician would have made him alert to signs which might have resulted in the initiation of prophylactic measures For example charts of patients who died from pulmonary embolism have contained notes such as "patient complains of pain in left calf" or "pain in the right groin" or "sudden breathlessness" or a "sharp pain in the right chest" Many other examples could be cited Because these initial signals were of short duration disappearing within a few days or even hours their sinister significance was not recognized and steps were not taken to prevent further development of thromboembolic complications As Barnes pointed out 15 years ago little could then be done except wishful thinking if the pulmonary emboli were arising from the heart If they were arising from the veins of the legs ligation proximal to the site of a recognized thrombophlebitis appeared logical and was the favored method of attack Experience has demonstrated however that 1 ligation does not prevent the active thrombophlebitic process from continuing 2 emboli may and frequently do continue to arise from veins other than those ligated 3 fatal emboli may arise from thrombi which have formed immediately proximal to the site of ligation even though this may be as high as the inferior vena cava 4 the late effects of ligation of the femoral or iliac veins or the inferior vena cava are often undesirable with serious evidence of venous insufficiency associated with pain edema and ulceration Therefore ligation was found not to be a preventive measure of choice except when it was desirable to eliminate varicose veins a common site for recurrent thrombophlebitis with secondary emboli

Erb and Schumann (7) reported their experience in 100 cases of fracture of the femur a condition very frequently complicated by pulmonary embolism Fifty patients were subjected to bilateral femoral vein ligation and 50 were used as controls The mortality rate was not lowered in the surgical group indeed the ligation itself was frequently followed by thrombosis in the site proximal to

the ligature Pulmonary embolism was increased

That surgical interruption of veins is not a treatment for phlebitis is well illustrated by a patient of ours While in the army he had a sudden pulmonary embolus Examination disclosed phlebitis in his left leg A left superficial femoral vein ligation was performed During the convalescence from this operation a second pulmonary embolus occurred Examination disclosed phlebitis in his right leg This time in an effort to trap all possible emboli his inferior vena cava was ligated Subsequently phlebitis developed in his arms and thereafter he had multiple pulmonary emboli Since his physician did not wish to tie off the superior vena cava he was referred to us for long term anticoagulant therapy There have been no further emboli during a period of eight years

The advent of anticoagulant drugs permitted a new approach to the problem which is actually based on the fact that under certain conditions the blood clots too easily The mechanism for this easy clotting is not understood Exhaustive studies of the clotting factors frequently fail to clarify the picture Pieces of the original clot break off and travel to the lungs If the original clot could be prevented from forming no such problem would ever arise If such a clot once formed could be contained as a small process and allowed to seal itself off instead of propagating a tail which could break loose the risk would be markedly lessened If emboli once delivered could be prevented from propagating and blocking off more branches of the pulmonary arterial tree the risk to life would decrease All of these accomplishments are theoretically possible with the perfect anticoagulant used correctly under favorable conditions All of them have been achieved in varying degree and often with striking success with the presently available anticoagulants Many studies have demonstrated a striking reduction in mortality and morbidity when anticoagulants have been used either prophylactically prior to the first pulmonary embolus or even after the first embolus in the prevention of subsequent ones

A summary of the results of several large series of patients with postoperative thrombophlebitis shows the following once thrombophlebitis has been recognized in the absence of anticoagulant therapy the risk of pulmonary embolism ranges from 4 to 60 per cent Of those patients who have had one embolus the risk of death from a subsequent embolus is approximately 20 per cent In contrast patients with postoperative thrombophlebitis who receive adequate anticoagulant therapy suffer from pulmonary emboli in less than 5 per cent of cases and the mortality rates are under 0 5 per cent (8) These are indeed striking figures They have been duplicated with heparin heparin and the coumarin derivatives combined and the coumarin derivatives alone In obstetric cases (9) the incidence of pulmonary embolism following untreated thrombophlebitis has been reported as ranging from 15 to 35 per cent with a mortality rate of 3 to 5 per cent This low mortality may be associated with the finding of the Committee on Anticoagulants (10) that while the younger patients had nearly as many emboli as older patients the mortality rate was much lower probably because of their ability to stand the reaction or shock In addition it is probable that more of the pulmonary emboli in obstetric patients arise from pelvic veins of smaller caliber and hence are less likely to be fatal

While it appears that once a thrombus or a pulmonary embolus has been recognized anticoagulant therapy is the most important form of therapy certain other measures should be borne in mind

As mentioned previously when carrying on a physical examination of the chest the physician should scrupulously avoid having the patient inhale deeply or cough since such maneuvers increase the negative venous pres

sure thus developing a most favorable condition to suck loose the tail of a clot which may then travel into the lungs Deaths have occurred directly following such maneuvers The patient must be warned against straining at stool thus producing a Valsalva experiment following which the hemodynamic pressures are markedly disturbed In one series 20 per cent of the deaths from pulmonary embolism were reported as occurring at stool The physician must provide laxatives to prevent this situation

As discussed elsewhere the acute reaction of pulmonary embolism has been thought to be due in part to spasm of the artery This led to the use of antispasmodic substances including papaverine aminophylline atropine and opium derivatives separately or in combination as for example Spasmalgin Favorable results have been reported following their use but large well controlled series are not available Nevertheless it seems advisable to use these or other relaxing agents to allay the extreme tension so commonly present in these patients

Twenty years ago serious attempts were made to develop operative procedures for removal of pulmonary emboli but they were very hazardous and with the advent of anticoagulant therapy they have been abandoned The use of oxygen is indicated in the presence of dyspnea or cyanosis In recent years efforts have been made to reduce the incidence of the postoperative thromboembolism prior to its original development The most popular methods have included the following

1 Early ambulation within 24 hours of the operation or delivery is still a matter of controversy but we incline toward the evidence in favor of it Active proponents of this method have pointed out that merely sitting in a chair as commonly practiced is not ambulation and indeed greater stasis may occur with the legs in dependency for long periods than if they are actively moved in bed or by walking In all patients we prefer the oscillating bed

2 Deep breathing many times a day should be immediately discontinued on any sign of thrombosis or embolism

3 In the treatment of phlebitis we elevate the foot of the bed to speed up the flow of blood through the venous channels of the legs and pelvis and to prevent stasis For many years it has been our practice to do this as a prophylactic measure following surgery whenever the surgeon feels the operative procedure permits it and also provided that the arterial supply to the limbs is adequate (15) None of these surgical patients has had a postoperative pulmonary embolus Torpin (12) has reported similar results namely freedom from pulmonary emboli following 1,500 gynecologic operations using this simple prophylactic measure The use of an oscillating bed for chronically ill or postoperative patients has much to recommend it The repeated filling and emptying of the veins should aid in prevention of phlebitis

4 Elastic stockings are used in the prevention of pulmonary embolism Wilkins and Stanton (16) have recently reported that in a series of over 5 000 routine hospital admissions pulmonary emboli were significantly reduced by the continuous wearing of light elastic hose

5 Prophylactic ligation of the veins postoperatively is much less popular than it was 10 years ago Some workers have even reported more emboli after ligation than in a control series We do not favor this procedure

6 We should not recommend anticoagulant therapy on a purely prophylactic basis in all patients or after all operations but certain situations probably justify its use according to large series reported by Barker Brambel and others and our own experience These include 1 major pelvic surgery 2 herniorrhaphy 3 major abdominal surgery especially in the presence of a history of former thromboembolic conditions 4 surgery on older

persons 5 surgery in the presence of auricular fibrillation, coronary artery disease or passive congestion, 6 the presence of marked varicose veins or venous insufficiency 7 after delivery when the mother has had previous thrombophlebitis or pulmonary embolism and 8 in all patients who have previously demonstrated a tendency toward thrombosis or embolism

Long Term Anticoagulant Therapy The treatment of pulmonary embolism should not be limited to a consideration of the acute episode first encountered This may be a single experience secondary to thrombosis which becomes organized and causes no further trouble and treatment of three to four weeks duration may be sufficient On the other hand the patient may continue to have repeated emboli from chronic recurrent thrombophlebitis or from a heart in auricular fibrillation We have patients who have had as many as twenty emboli from such sources It then becomes necessary to employ long term anticoagulant therapy The results of such therapy are encouraging and have been reported by us (13 14) We have now maintained more than 100 patients for from 1 to 13 years on anticoagulants

Case 21 Recurrent Idiopathic Phlebitis (Long Term Anticoagulant Therapy)

Mrs B has a family history of intravascular clotting Her father died of a pulmonary embolus Her mother had had recurrent phlebitis and also died of a pulmonary embolus

At the age of 42 she developed a superficial phlebitis which extended to the deep veins A pulmonary embolus followed This was before the era of anticoagulants so both femoral veins were ligated to prevent further emboli During the next several years she developed repeated attacks of phlebitis in the legs and arms Each attack resulted in invalidism Dicumarol therapy was started and continued successfully for eight years Attempts have since been made to take her off treatment but as the prothrombin time sinks to a normal range fresh phlebitic areas appear

There have been no ill effects from her long term anticoagulant therapy nor has there been any serious bleeding At one time she commenced to have increased menstrual bleeding This is unusual with a normal uterus and in properly administered anticoagulant therapy Papanicolaou smears were positive A hysterectomy was performed and a small carcinoma was found The anticoagulant therapy had led to its bleeding and early discovery

This patient has resumed her dicumarol treatment She has now been on this regimen nine years and the clotting tendency in her blood although not understood may necessitate continuation indefinitely of anticoagulant therapy

Case 22 Carcinoma of Breast with Multiple Vascular Thrombi



Embolus occluding renal vein



Microscopic section of lung with embolus

This young woman with a rapidly growing carcinoma of the breast developed multiple thrombi in her veins heart and arteries

The left figure shows the kidney and renal vessels Note the occlusion of the renal vein by an antemortem clot

The right figure is a microscropic section of lung Note the embolus present in the pulmonary artery

Case 23 Pulmonary Emboli Causing Right Heart Failure



A Note large dilated right ventricle and the embolus in pulmonary artery



B Microscopic section of lung with embolus

This 48 year old housewife was admitted to the hospital on December 19 1955 because of dyspnea on exertion that had become progressively worse over the past 10 years

HISTORY The patient was so active as a child that she was considered a tomboy When she was 10 years old tuberculous cervical lymph nodes were removed at another hospital She made a good recovery with no apparent after effects At the ages of 25 and 27 she became pregnant and had no difficulty during the birth of her children No history of rheumatic fever or chorea was obtained In 1945 dyspnea on exertion gradually developed while she was living in Reno Nevada An x ray unit visited her neighborhood in 1953 and the patient had a chest x ray taken She was told to visit her physician because the film showed an abnormal chest finding She ignored this advice but when the dyspnea became worse and orthopnea became prominent she did visit her doctor in June 1955 Chest pain hemoptysis and dependent edema had not occurred but she had gained 20 pounds in the past two years and weighed 165 pounds Both the physicians x rays and the mobile unit film showed bilaterally enlarged hilar shadows Bronchoscopy with bronchial washing studies showed no abnormalities A supraclavicular node biopsy was also done but the results are not available Because of the progression of her symptoms she came East and entered The New York Hospital on December 19 1955

The temperature pulse and respirations were normal The patient was a well developed and slightly obese woman who did not appear in any distress Healed scars were present on both sides of the neck and there was a more recently healed left supraclavicular

scar The neck veins were not distended and there was no dependent edema There was no clubbing of the fingers or toes A few scattered rales were heard at the bases of each lung The heart was slightly enlarged to the right and left A mild heave was felt over the region of the right ventricle and the P_2 was palpable The pulmonic second sound was accentuated and ringing A very faint systolic murmur was heard over the pulmonic area and a moderate high pitched diminuendo diastolic murmur was noted along the left sternal border at the second and third left intercostal spaces No other abnormalities were found on physical examination The urine was normal and the hemoglobin was 16 3 gm per cent red blood cell count 5 3 million hematocrit 49 per cent and white blood cell count 8 400 with a normal differential The Mazzini test showed a doubtful positive reaction The blood urea nitrogen was 12 mg per cent and the CO_2 was 21 mM/L A chest film showed moderate cardiac enlargement and abnormal prominence of the pulmonary artery segment and right ventricle Some pulmonary congestion was also found An angiocardiogram showed some dilation of the right atrium and right ventricle with marked enlargement of the pulmonary artery and its major branches This study showed no evidence of a right to left shunt

Cardiac catheterization was done but a complete study was not possible because of the patient s lack of cooperation An elevated right ventricle pressure of 128/12 mm of Hg was found and the arterial oxyhemoglobin saturation was 85 3 per cent Simultaneous determination of oxyhemoglobin saturation from the right brachial artery was 87 9 per cent and from the right femoral artery 87 7 per cent The pH of the blood was 7 4 plasma CO content was 20 mM/L and CO tension 32 mm Hg The electrocardiogram showed an incomplete right bundle branch block with right ventricular hypertrophy She was digitalized and on January 4 1956 she left the hospital 16 days after entry and returned to Nevada

During the subsequent 21 months the dyspnea and orthopnea became worse and the patient lost 55 pounds During this period she had been hospitalized eight times in the West and was treated with mercurials digitalis phlebotomies and on one occasion with anticoagulants In July 1957 dependent edema and cyanosis became noticeable A hemorrhoidectomy was done in August 1957 at a Nevada hospital Because of her continued downhill course she reentered The New York Hospital September 29 1957

Examination The temperature was 37 C the pulse rate 100 respiration was 26 and the blood pressure 112/76 The patient was a thin cyanotic white woman who appeared chronically ill The lips nails tongue and buccal mucosa were cyanotic Slight clubbing of the fingers was found The neck veins were moderately distended and sacral edema was prominent The thorax was slightly increased in the anteroposterior diameter and scattered fine rales at both bases were heard The heart was enlarged 11 cm to the left of the midsternal line in the fifth intercostal space A broad heave was felt over the sternum The pulmonic second sound was accentuated A blowing systolic murmur was heard at the apex At the third interspace along the left sternal border a soft blowing systolic murmur and a diminuendo diastolic murmur were described The abdomen was obese and the liver was felt down to the umbilicus but the spleen was not felt

Laboratory Data Urine protein 1+ was noted and the sediment contained many clumps of white blood cells Blood the hemoglobin was 16 5 gm per cent the red blood cell count was 5 1 million the hematocrit was 57 per cent the white blood cell count was 13 200 with 59 polymorphonuclears 11 bands 21 lymphocytes 8 monocytes and 1 eosinophil the platelets were adequate The Mazzini test was again doubtfully positive The blood

urea nitrogen was 12 mg per cent bilirubin 0 7 mg per cent and uric acid 3 8 mg per cent carbon dioxide 24 mEq/L chlorides 98 mEq/L sodium 0 135 mEq/L and potassium 4 8 mEq/L The prothrombin time was normal X ray the chest showed an increase in cardiac size as compared with films of the first admission Enlargement of chambers and pulmonary artery was the same as that found in the past Electrocardiogram this showed a NSR with the incomplete right bundle branch block and right ventricular hypertrophy Stool guaiac negative

COURSE Shortly after admission a phlebotomy of 350 ml was done She was treated with mercurial diuretics and digitalis was maintained On the fourteenth day she fell from her chair and struck the right side of her head but did not suffer any laceration or lose consciousness A skull x ray showed no fracture A second phlebotomy of 300 ml on the twelfth day failed to lower the hematocrit which remained at 56 per cent The patient was a nursing problem refused to take food or medications and was very apprehensive Respirations became progressively more labored and she died on the seventeenth day

AUTOPSY FINDINGS The autopsy revealed massive old and recent pulmonary thromboemboli together with extensive sclerosis of the large and small arteries and arterioles right ventricle hypertrophy and dilatation No congenital anomalies were found in the heart The changes in the pulmonary vessels apparently were the result of repeated old and recent embolization with subsequent thrombosis

Figure A (p 51) shows the profound hypertrophy and dilatation of the right ventricle enlargement of the pulmonary artery and emboli present in the lumen

Figure B (p 51) shows extensive thickening of the wall of pulmonary arterioles and a small artery

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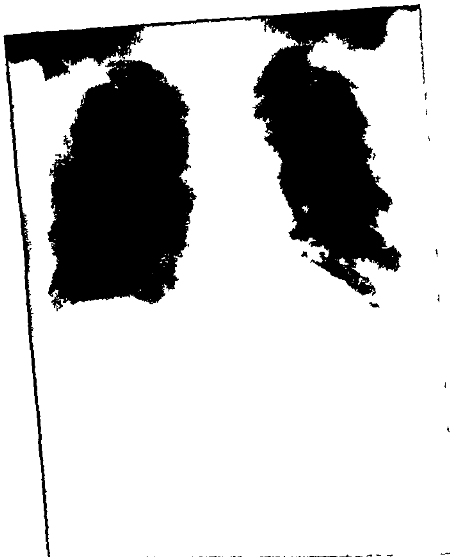
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Case 25 Acute Pulmonary Infarction



A 50 year old man (The New York Hospital #625583) had a cholecystectomy performed because of cholelithiasis On the eighth postoperative day he experienced sharp pain in the right chest and raised blood streaked sputum Deep phlebitis was found in his right calf Anticoagulants were administered promptly the leg was wrapped in warm moist packs and elevated An uneventful recovery ensued

The illustration shows a portable chest film The right diaphragm is elevated and tented A pleural effusion is present There is partial atelectasis of the right lower lobe

Case 24 Multiple Small Pulmonary Emboli

A 57 year old social service administrator (The New York Hospital #530436) was first seen in November 1948 During the previous 10 years she had had eight illnesses that had been diagnosed as lobar pneumonia On admission she complained of cough chest pain and blood tinged sputum Examination showed typical signs of mitral stenosis X ray of the chest showed a shadow in the right lower lung field Our diagnosis was pulmonary infarction secondary to an embolus from rheumatic heart disease Dicumarol therapy was instituted and maintained for two months The patient lapsed from treatment Again she had a sudden episode of chest pain cough and blood streaked sputum



Note generalized cardiac enlargement, bilateral pleural effusion and increased density of the posterior basal segment of the right lower lobe at site of pulmonary infarction

Case 26 Pulmonary Emboli

A 51 year old woman (The New York Hospital #570625) was first seen in June 1950 with a pulmonary infarction At the age of 14 she had had rheumatic fever and at eighteen she had been told she had a heart murmur Until her present illness she had been entirely well She was a business executive and led a busy professional and social life The day prior to admission she had developed severe pain in the right lower chest radiating to the back Examination disclosed signs of mitral stenosis and aortic stenosis There was a regular sinus rhythm interspersed with many premature contractions Examination of the chest revealed dullness and diminished breath sounds at the right base posteriorly

X ray of the chest is shown in the accompanying illustration This film shows generalized cardiac enlargement bilateral pleural effusion and an increased density in the posterior basal segment of the right lower lobe which is consistent with the clinical findings of pulmonary infarction

The patient ran a febrile course for a week She was given anticoagulants starting with heparin and shifting to dicumarol She was digitalized and discharged on her fourteenth hospital day Dicumarol was discontinued

Two weeks later a second pulmonary infarction occurred She was readmitted to the hospital and again given dicumarol The decision was made to keep her on anticoagulants permanently There have been no further emboli during a period of six years She reports every 14 days for prothrombin determinations She has had several minor hemorrhagic manifestations in the form of easy bruising after slight trauma but no major ones Her prothrombin time varies from 20 to a high of 40 seconds Her average daily dose is 75 mg of dicumarol While under treatment she has made two business trips to California one to Paris and Rome and many shorter trips Her blood tests have been arranged at various points with physicians interested and trained in this work Her course was complicated four years ago by the development of hyperthyroidism This was successfully controlled with radioactive iodine

This patient represented a case of rheumatic heart disease not fibrillating in which emboli from the right heart were delivered to the lungs

Case 28 Pulmonary Embolism



Radiograph of pulmonary embolism



Note localized area of infiltration in right posterior costophrenic sulcus with pleural reaction

Examination showed splinting of the chest on the right side Her temperature rose to 39 C Examination of the legs was completely normal The white blood count was 7 800 the differential count showed 69 mature polymorphonuclear leukocytes 10 bands 12 lymphocytes 8 monocytes and 1 eosinophil

Moist rales were present at the posterior base on the right side X ray of the chest (shown in the accompanying figures) showed a localized area of infiltration in the right posterior costophrenic sulcus with a small amount of pleural reaction around it

In the differential diagnosis the conditions to be considered were a pneumonitis or a pulmonary embolus The time of onset of pleuritic pain and the response to therapy made the diagnosis of pulmonary embolus most likely During any extensive surgery in the pelvis a great many small veins have to be tied off These veins thrombose and thus represent potential sources of emboli Whether or not this is of clinical significance depends on whether or not a clinically detectable embolus actually does occur

The only therapy consisted of the use of anticoagulants In this case these were administered as follows heparin (concentrated to 20 000 units per ml) was given subcutaneously every 12 hours an initial dose of 15 000 units was given Blood coagulation tests were done daily The aim was to obtain a maximum of three times the control coagulation time in a period of four hours after administration of the heparin Coagulation time should return approximately to normal by the twelfth hour The dose was varied each day ranging from a low of 10 000 units to a high of 15 000 units This was kept up for a period of three weeks Heparin alone was used in this patient because her husband a distinguished cardiologist had a personal preference for it and urged us to do so The fever slowly decreased during this period Chest symptoms persisted for about four days and then gradually subsided Rales persisted for 12 days The abdomen was tender but not more so than one would expect from such a major surgical procedure The legs were at all times clinically normal Because of the absence of involvement in the legs it was not necessary for this patient to wear elastic stockings

Case 27 Pulmonary Embolus



Note increased vascular markings and bilateral pleural effusion



Four and a half seconds after intravenous injection of contrast medium

A 44 year old white man had had three previous episodes of myocardial infarction He also had had multiple pulmonary infarctions The x ray of the chest (see the left figure) shows increased vascular markings in both lung fields and a bilateral pleural effusion The angiocardiogram (see the right figure) by Dr Israel Steinberg made four and a half seconds after the intravenous injection of the contrast medium shows the pulmonary conus pulmonary artery and its major branches well outlined The branches to th right lower lobe are irregular and cut off and indicate the point of lodgment of a pulmonary embolus

The patient died several months later Au topsy showed a large embolus in the righ lower pulmonary artery branch multiple ol pulmonary infarctions and multiple myocard al infarctions He had not received antico agulants

Case 28 Pulmonary Embolism following Pelvic Thrombophlebitis

Mrs D P (The New York Hospital #711090) age 65 a housewife was found to have a carcinoma of the endometrium Operation was performed on January 28 1957 It consisted of total removal of the uterus the tubes the ovaries the lymph nodes in the iliac and obturator areas Pathologic diagnosis was found to be adeno acanthoma with slight to moderate endometrial invasion and vascular lymphatic involvement

The patient had a very calm postoperative course during the first 12 days Her temperature remained between 37 and 38 C she ate well and her morale was good She was allowed up to the bathroom had her meals sitting up and walked about the room On February 10 eleven days after the operation she complained of pain in the right lower chest The pain was made worse by lying on the right side Deep breathing was impossible

One second film



Three second film





Six second film



Fourteen second film

Case 29 Pulmonary Embolus, Angiocardiogram



Front view showing en larged pulmonary conus

This young woman with rheumatic heart disease auricular fibrillation and mitral sten osis had a large pulmonary infarction

The above photograph shows a front view of the chest The pulmonary conus is enlarged The right diaphragm is prominent a pleural effusion is present and the right lower lobe is obscured by a mottled shadow

The photograph on the facing page shows four views mounted on one slide of the angio cardiogram by Dr Israel Steinberg at intervals of 1 3 6 and 14 seconds after the intravenous injection of contrast media *The 1 second film* shows the contrast media outlining the superior vena cava and right atrium *The 3 second film* shows the pulmonary conus and pulmonary artery branches well outlined They are greatly dilated *The 6 second film* shows ar borization of the pulmonary branches with sharp termination of a large branch to the right lower lobe probably at the site of the embolus The left auricle is outlined and shows a double density possibly the site of a mural thrombus *The 14 second film* shows the left ventricle and aorta outlined Note how small the aorta is compared to the dilated pul monary artery

SUMMARY

The importance and frequency of pulmonary embolism have been discussed. The diagnosis and treatment have been presented. Important strides have been made in the treatment of this condition during the past 12 years as a result of the advent of anticoagulant drugs. While death and recurrent emboli may occur during such therapy, the incidence of both has been strikingly reduced by the use of heparin, coumarin and indanedione derivatives.

Greater awareness of the possibility of thrombosis and pulmonary embolism results in earlier diagnosis and treatment and further reduction in mortality from these conditions.

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Case 30 Pulmonary Embolism, Migratory Phlebitis, Carcinoma of Pancreas

Certain types of carcinoma seem to derange the blood clotting mechanism and give rise to intravascular clots Perhaps carcinoma of the pancreas is the most apt to do this as was pointed out by Trousseau nearly a century ago Carcinoma of the lung stomach breast and metastases to the liver are also frequently associated with clotting

Case 30 was a middle aged man who developed phlebitis and pulmonary embolus while crossing the Atlantic on a ship The condition was quickly controlled by anticoagulants when he arrived in New York

Phlebitis recurred while receiving therapeutic doses of dicumarol This is characteristic of phlebitis associated with carcinoma However it does respond to parenteral heparin We were so convinced that this patient had a hidden carcinoma that we urged a laparotomy even though all diagnostic chemical and x ray procedures were essentially normal

At operation a carcinoma of the tail of the pancreas with extension to the liver and peritoneum was found

3

Lymphedema

Lymphedema often presents a perplexing problem in diagnosis and management. The diagnosis is often confused with cardiac and renal disease. Treatment may be thereby misdirected or delayed and the condition worsens often irrevocably. The purpose of this short chapter is to assist in the early recognition and proper therapy of lymphatic disease.

TREATMENT OF LYMPHEDEMA

This depends upon complete cooperation between physician and patient in applying physiologic and psychologic principles over long periods of time. It requires the application of methods which are often uncomfortable and tedious.

The physician should maintain a degree of optimism and patience which will test him in the face of little or very slow improvement. At times rapid improvement occurs, especially after recent thrombophlebitis and in some cases which involve the arm. However, in lymphedema of the leg the objective may resolve into simply preventing a worsening of the condition. In many patients without proper therapy the edema progresses to elephantine size.

CLASSIFICATION

The 1953 edition of the Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Blood Vessels (8) defines and classifies lymphedema as follows:

Lymphedema. This refers to edema resulting from obstruction of the lymphatic flow. Microscopically, the early changes consist of dilatation of the lymphatics and widening of the tissue spaces due to edema. In longstanding lymphedema there is proliferation of the connective tissue, often a variable degree of inflammatory cell infiltration, pigmentation and fibrotic thickening of the dilated lymphatics.

Clinically, the changes can be correlated with the fact that at first the edematous tissue is compressible (soft edema) but later the swollen tissue is firm (so-called brawny induration or hard edema). It is characteristically pale in contrast to the cyanosis or rubor seen with venous stasis. The term elephantiasis is used for the grotesque deformities of the more severe types. Lymphedema may be primary or secondary, but the former is better designated as idiopathic.

Clinical Features

Table I Differential diagnostic features

	Lymphedema	Thrombophlebitis
Color of leg (horizontal position)	Pale	Usually slightly cyanotic
Veins	Usually hidden by edema	Collateral veins often prominent If patient has had previous attacks varices may be present as well as plevi usually around the ankle
Pigment deposits	None	In presence of long standing venous insufficiency there is often much brown hemosiderin deposit around ankles and shins
Inflammation	Cellulitis is usually diffuse and often bright red	Red hot tender venous cords are pathognomonic but with deep thromboses these may not be discernible
Tenderness	None except in presence of cellulitis	Marked tenderness to deep pressure Homan's sign is sometimes present if deep calf veins are involved

The etiology of edema of an extremity following surgery is a controversial subject (3) Cancer may invade and block the lymphatic depots The surgical and radiologic destruction of main lymph channels often leaves insufficient patent vessels for adequate drainage Fluid accumulates in the tissue spaces In time there is a connective tissue proliferation that converts the boggy areas into a fibrotic mass

The most common form of swollen leg we are called on to treat is that of lymphedema praecox This condition usually occurs in young women rarely in men The ratio is about 10 to 1 but the cause for this difference is not understood It develops most frequently shortly after the onset of the menses Occasionally it is first manifested as late as 40 years of age The later it appears the milder it is apt to be although some early cases do not progress to a serious degree

Characteristically edema starts in one foot and ankle At first only a slight puffiness is present Gradually it increases spreading up the leg sometimes involving tissues of the thighs and buttocks It may be unilateral for many years before starting in the other leg In the beginning the edema is reduced during the night and develops during the waking hours when the patient is up Any constricting garment such as circular garters or girdles causes it to increase peripherally Girdles are particularly harmful for they are designed for women in the upright posture only When women sit the girdle stretches exerting pressure across the groin and it acts as a tourniquet to impede lymph flow Sitting for many hours in vehicles theaters, or at card playing sessions is a strain on even a normal lymph system

Good lymph flow requires body movement During active sports or walking the edema tends to decrease Walking in deep water produces the unique situation of a maximum compression pressure at the foot decreasing progressively up the leg When this is combined with the pumping action of muscular motion it presents an ideal method of squeezing fluid out of the leg Swimming is the best sport for these patients

The etiology of lymphedema praecox is completely obscure Clinically it seems as if there were a congenital blockage of the lymph channels or simply that the total lymph drainage system is not efficient enough Kinmonth and Taylor (6) have shown that the lymph trunks are dilated and the valves defective Because of the frequent episodes of cellulitis the condition is often confused with thrombophlebitis The table of differential points (Table 1) may be helpful in establishing a diagnosis (5) A familial lymphedema (Milroy's disease) develops in a similarly inexplicable manner This may involve one or more extremities or half the body

Classification of Primary and Secondary Lymphedema

A Primary or idiopathic lymphedema

1 Congenital (Milroy s disease) This is a hereditary congenital nonpainful lymph edema involving all or part of one or two extremities It may be unilateral involving the upper and lower extremities and rarely additional areas may be involved

2 Praecox This refers to a clinical syndrome affecting young females much more often than males The lymphedema usually appears between the ages of 15 and 35 involving first the feet later ascending to involve the legs and thighs

B Secondary lymphedema is due to

1 Surgical removal of lymph nodes

2 Neoplastic invasion of lymph nodes either by primary or metastatic neoplasm

3 Lymphadenitis following

a X ray treatment Fibrosis and scarring enhanced by x ray may cause obstruction of the lymphatics and resultant edema

b Pyogenic infection

c Granulomas due to

(1) Filariasis

(2) Lymphogranuloma venereum

(3) Tuberculosis

(4) Syphilis

4 Dependent edema Edema of the legs may follow prolonged sitting as during long trips by auto rail or air It has been reported in persons seated for many hours in air raid shelters Some complicating factor is usually present such as a girdle which folds and binds the groin when seated or a chair with a firm edge that presses on the thighs If the edema is pale the obstruction is lymphatic If it is cyanotic a complicating phlebitis may be present

CLINICAL FEATURES

The most common form of lymphedema of the arm follows radical mastectomy A study of this subject was published in 1951 (3) This type of lymphedema is characteristically a progressive disabling and often painful condition In some cases the swelling occurs shortly after operation in others edema formation may not develop for as long as five years It usually appears first in the hand and accumulates proximally although in some of our patients the reverse has been true The increased weight of the arm pulls on the nerve roots causing a neuralgia The pain often disappears when the swelling is reduced by treatment

Swollen arms and legs are aesthetically disturbing They may be devastating to the ego of otherwise well adjusted women Several of our patients have been recluses until reduction in swelling encouraged them to abandon their seclusion The deformity has resulted not infrequently in marital difficulties Even minor degrees of swelling may be sufficient to keep women from engaging in activities requiring a costume that reveals the abnormality

Edematous extremities are very often the site of attacks of cellulitis and erysipelas We first saw one patient in his one hundred and fourth attack which had occurred over a 31 year span He had kept records of his attacks in numerous hospital stays It is apparent from this clinical observation that tissue immunity is reduced in those areas with slow lymph flow The fact that lymph flow is slow is readily demonstrated by the intracutaneous injection of dyes in the manner described by McMaster (7) A simple furuncle that is quickly healed in a normal arm may become a large spreading infection in one with lymphedema Fungous infections splitting the skin permit infections to enter and are frequent complicating factors They must be rigorously controlled Elevation moist heat and appropriate antimicrobial drugs are usually quickly curative

quire a garter belt to hold them up They are strong enough to give adequate support for mild or moderate cases but still light enough not to buckle

The elastic sleeve which was described previously (3 4) has been improved by use of the new orlon material The sleeve should start at the base of the fingers and should have an opening for the thumb large enough to give it full play A pressure of 40 mm of mercury at the distal end of the sleeve gradually falling to 15 mm at its termination in the upper one third of the arm is adequate compression See the accompanying illustration Sometimes the fingers become very swollen A rubber glove worn with the sleeve may help to correct this

Physiotherapy Massage in the direction of the lymph flow toward the heart increases the speed of flow We recommend effleurage each evening for 10 minutes self administered While in the hospital massage may be performed more frequently

The Jobst Company together with Brush and Heidt (12) have developed a tight sleeve lined with inflatable balloons The balloons are alternately inflated by a pump which works on an automatic device We have found it of help in milking out" the fluid in lymph edematous arms and legs Although this may be a temporary gain it aids in promoting the efficiency of lymph flow

Hydrotherapy in the form of walking in deep water is of great value in forcing edema fluid from legs and has already been mentioned under Clinical Features"

Dehydration The disappearance of edema fluid can sometimes be hastened by a low salt diet and daily diuretics We employ a 2 gm



Foley elastic sleeve

salt diet and daily injection of 1 to 2 ml of thiomerin Fluids are not restricted This regimen is maintained only during the first week of hospitalization

Activity Patients are encouraged to resume full activity and sports wearing their elastic stocking or sleeve The pumping action of muscular movement on venous return and lymph flow is well known

Additional measures have been suggested as follows

1 Buried suture technic of Handley (5 9 10 11)
2 Hyaluronidase (1 2 11 13)
3 Sympathectomy

In our experience all three have failed to produce improvement and sympathectomy often makes the edema worse It is definitely contraindicated

MEDICAL MANAGEMENT

The judicious application of sound physiologic principles is the essence of treatment of lymphedema

Gravity Edema of the arms or legs is invariably reduced by prolonged elevation which as is readily demonstrated reduces venous pressure and increases the speed of lymph flow Elevation of the lower extremities is easily accomplished by placing blocks under the bed posts at the foot Six to eight inches is the maximum elevation that is well tolerated Raising the entire foot of the bed in this manner is much more comfortable and far more practical than either raising the mattress alone or placing pillows under the legs The *new type wedges placed under the lower half* of the mattress are apt to produce low back strain unless the patient sleeps constantly on *his back Patients who sleep with their legs* properly elevated receive good lymph drainage in the legs for at least one third of each day

Sleeping with the arm in maximum elevation is a much more difficult problem After a number of years of experimentation with various measures and methods an apparatus was designed to maintain the arm in maximum elevation and to allow the patient to move about in bed as well as from side to side (see illustration for Case 34) This is primarily intended for patients in whom the swelling represents a serious situation This apparatus uses the principle of traction with enough counter weights to balance the arm exactly It is most important that no constricting bandages be *used to attach the arm to the traction apparatus* Under tension these bandages will act as tourniquets Instead foam rubber pads are loosely applied to the arm by an elastic bandage The friction of the foam rubber against the skin is enough to secure it A halter snap allows the patient to disengage herself quickly from the apparatus

Before placing any limb in elevation one must be certain that the arterial supply is adequate In the presence of arterial insufficiency grave damage may be done by elevation of the part If the radial or pedal pulses are present and pallor does not develop in the toes or the fingers on elevation it may be assumed that adequate arterial flow is present Interference *with the radial pulse is sometimes* caused by changing the position of the arm This pulse should be palpated in the position of extension to be sure of its quality (test for hyperabduction and scalenus syndromes)

Most frequently satisfactory reduction in the size of swollen arms can be obtained in women who have consented to enter the hospital for the inception of therapy There maximum elevation can be applied without interruption Patients learn the therapy and cooperate more fully than if they are simply given instructions to carry out at *home* If however they are unable or unwilling to continue *their regimen at home* they may lose a part or all of their improvement

Compression Edema fluid drains out of the extremities in elevation *but it tends to* reaccumulate in dependency To prevent this elastic stockings should be worn on the legs or an elastic sleeve on the arm when the patient is up and about

The stocking should be made to measure It should fit snugly and start just above the toes The foot area should give a compression of 40 mm of mercury The pressure should gradually fall as the stocking progresses up the leg In most cases the stocking may end just below the bend of the knee When edema of the thigh is also present the stocking should continue nearly to the groin

Most long stockings tend to buckle in back of the knee giving a tourniquet effect in the popliteal space on walking or especially on sitting *Recently orlon stockings have appeared* on the market Long ones of this material re

quire a garter *belt to hold them up* They are strong enough to give adequate support for *mild or moderate cases but still* light enough not to buckle

The elastic sleeve which was described previously (3 4) has been improved by use of the new orlon material The sleeve should start at the base of the fingers and should have an opening for the *thumb large enough* to give it full play A pressure of 40 mm of mercury at the *distal end of the sleeve* gradually falling to 15 mm at its termination in *the upper one third of the arm* is adequate compression See the accompanying illustration *Sometimes the fingers* become very swollen A rubber glove worn with the sleeve *may help to correct this*

Physiotherapy Massage in the direction *of the lymph flow toward the heart increases* the speed of flow We recommend effleurage *each evening for 10 minutes* self administered While in the hospital massage may be per *formed more frequently*

The Jobst Company together with Brush and *Heidt (12) have developed a tight sleeve* lined with inflatable balloons The balloons are *alternately inflated by a pump which* works on an automatic device We have found it of help in milking out" the fluid in lymph edematous arms and legs Although this may be a temporary gain it aids in promoting the efficiency of lymph flow

Hydrotherapy in the form of *walking in* deep water is of great value in forcing edema fluid from legs and has already been mentioned under Clinical Features

Dehydration The disappearance of edema fluid can sometimes be hastened by a low salt diet and daily diuretics We employ a 2 gm



Foley elastic sleeve

salt diet and daily injection of 1 to 2 ml of thiomerin Fluids are not restricted This regimen is maintained only during the first week of hospitalization

Activity Patients are encouraged to resume full activity and sports wearing their elastic stocking or sleeve The pumping action of muscular movement on venous return and lymph flow is well known

Additional measures have been suggested as follows

1 Buried suture technic of Handley (5 9 10 11)
2 Hyaluronidase (1 2 11 13)
3 Sympathectomy

In our experience all three have failed to produce *improvement and* sympathectomy often makes the edema worse It is definitely contraindicated

Case 31 Massive Lymphedema of the Arm

Mrs L E is a woman of 59 years (The New York Hospital #647569) In 1942 a left radical mastectomy was performed for carcinoma This was followed by deep x ray therapy There was no wound infection The arm commenced to swell shortly thereafter

In one year it had reached a large size A Kondoleon operation was performed The swelling recurred and the following year a *second Kondoleon operation was performed* The arm gradually became larger When first seen by us on November 19 1951 it had reached a massive size (see the left figure) She had developed intense root pain in the neck and shoulder The arm was "too heavy to carry around She had become a recluse

She was hospitalized and given the regimen of elevation massage dehydration and compression outlined previously A very great improvement was obtained (see the right figure) in a period of 12 days (December 31 1951) There had been a great loss of edematous fluid leaving loose baggy skin (Table 2) The loose skin contracted slowly during the following six months The root pain had disappeared and she was discharged from the hospital with instructions to sleep with the arm elevated in the traction apparatus to wear the elastic sleeve during the day and to massage the arm each night for 10 minutes

This woman continues to follow our outline of therapy and has maintained the improvement The loose baggy skin contracted to the size of its contents The cosmetic appearance has improved greatly The former great weight of the arm was relieved and with it the pain from nerve root traction has disappeared Hyaluronidase failed to give further decrease in swelling

Table 2 Measurements of circumference in centimeters for Case 31

Location	Date 11 19 51	12 31 51	1 24 5?	2 27 5	4 2 53	2 2 54	6 19 56
Hand	28 0	23 8	23 5	21 6	22 5	19 7	19 8
Maximum Forearm	44 5	34 9	35 6	33 7	35 2	34 4	34 ?
Maximum Arm	52 1	39 9	37 8	38 2	37 1	36 8	36 7



Enormous swelling of the entire arm and hand



Twelve days later Patient was treated by elevation massage and Foley sleeve Swelling has largely subsided leaving bags of pendulous skin This loose skin shrank up slowly during the following six months

Case 32 Lymphedema of the Arm

Mrs. M. J. (The New York Hospital #426954) is aged 65 years. A left radical mastectomy was performed in November 1945 for canalicular *scirrhous carcinoma of the left* breast with no discoverable axillary metastases. Swelling of the arm developed following operation. It commenced first in the brachium. Slowly through the course of the next three years the swelling involved the lower arm, wrist and hand. The swelling usually subsided when the patient elevated her arm on a pillow, but during the two *months immediately preceding vascular consultation*, the swelling apparently became more fixed.

The patient was seen on July 8, 1948. She was a tiny woman and was very apprehensive and disturbed about the appearance of her swollen arm (see the top left figure). There was a large elliptical surgical scar across the left anterior thorax extending into the axilla. The axilla was deep; no nodes were palpable. Oscillometric readings were normal. The entire arm was swollen. The brachium and lower arm were indurated, and the swelling did not pit, although the hand and fingers had a soft edema which pitted readily. Measurements are shown in Table 3. Treatment, consisting of *nightly elevation, effleurage, and the use of* an elastic sleeve and a snugly fitting surgeon's rubber glove, was started at once.

Four weeks later the swelling had entirely cleared in the fingers and hand. The top right figure shows the Foley sleeve in place. There was marked reduction in the forearm and arm. The rubber glove was discarded. After six months the elastic sleeve was also eliminated. *On the patient's last visit, eight years after the* start of treatment, only a slight thickening remained in the forearm and arm (see the bottom figure). She continues to sleep with the arm elevated.

Table 3. Measurements of circumference in centimeters for Case 32

Location	Date 7-8-48	7-28-48	12-16-50
Hand (above metacarpal bulge)	18.5	18.1	18.0
Wrist	15.0	14.7	14.7
Maximum Forearm	25.0	20.0	19.7
Maximum Midbrachium	27.0	22.0	21.0

Close up of arms and hands



Foley sleeve in place





Eight year follow up

Case 33 Lymphedema of the Arm



Note swelling of hand and forearm



After treatment. Note wrinkling of skin and reappearance of veins.

Miss M. B. (The New York Hospital #553402) is aged 53. A radical left breast amputation was performed for carcinoma on October 14, 1949. One month later swelling developed in the arm. The area adjacent to the elbow became stiff and hard. In May 1950 the hand and fingers became swollen.

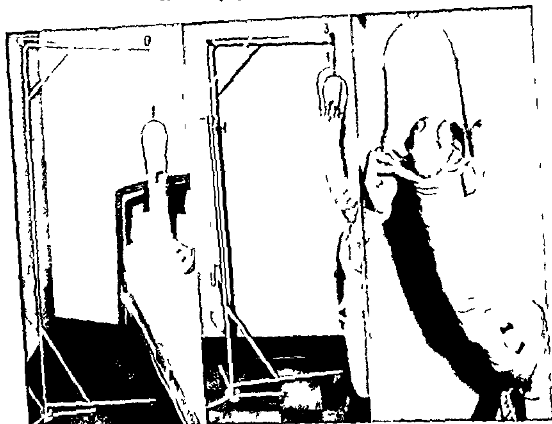
The patient appeared for vascular consultation on May 26, 1950. At that time the only abnormality was the surgical scar and the swollen arm. For a distance of 10 cm. above and below the elbow there was a woody dense edema. The arm was pale and firm. The upper arm was swollen but soft, while a very soft edema was present over the forearm, hand and fingers (see left illustration). Oscillometric studies were normal.

The patient was not hospitalized. She was instructed to suspend the arm in maximum elevation at night, wear an elastic sleeve, and massage the arm daily. The swelling left the fingers, hand and wrist rapidly (Table 4). The right illustration shows the wrinkling of the skin and the reappearance of the veins. The swelling has slowly subsided elsewhere. On the patient's last visit, October 10, 1954, the area around the elbow was soft.

Table 4 Measurements of circumference in centimeters for Case 33

Location	Date 5-26-50	10-18-50	10-10-54
Hand (above metacarpal bulge)	18.5	17.0	16.5
Wrist	15.5	15.0	15.0
Maximum Forearm	27.0	25.0	24.8
Maximum Brachium	28.4	28.0	27.0

Case 34 Lymphedema of the Arm



Use of the Foley arm edema stand

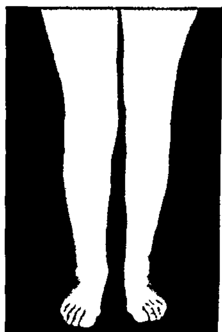
Mrs A G is a 57 year old Italian American housewife (The New York Hospital #493717) In 1947 a right radical mastectomy was performed for canalicular carcinoma of the breast with metastases to the axillary lymph nodes There was no wound infection She did not receive radiation therapy In 1950 the arm and hand became noticeably swollen Swelling gradually increased On February 27 1953 medical therapy was started Swelling decreased during the course of the first week especially in the upper arm where it was most marked

On March 9 and 10 nylon sutures were inserted One extended subcutaneously from the scapular region to the wrist the second from the supraclavicular area to the midarm Hyaluronidase was given daily for one week in high and low dosages directly into the edematous areas Only slight further improvement was noted in the course of the next six months At that time October 3 1954 the metastases were found in the cervical nodes

It is most interesting to observe that medical management improved the arm even with the development of metastases

The accompanying photographs illustrate the use of the Foley arm edema stand with the patient in various positions

Case 35 Lymphedema Praecox



Miss A McF (The New York Hospital #614483) a 23 year old stenographer was first seen on December 22 1950 She gave a history of having developed swelling in the right foot and leg in 1947 In June 1949 a right lumbar sympathectomy was performed in another hospital As has been repeatedly noted with other similar patients following this procedure the leg rapidly increased in size When first seen she had developed a massive edema extending from the waist to the toes on the right side With medical management the edema was held in check during the next eight months A paronychia developed in the right great toe She was admitted to The New York Hospital for surgical treatment of this infection and responded well While in the hospital in November 1951 two nylon sutures were implanted surgically by Dr William Terwilliger One extended from the medial side of the ankle to the groin and the second from the lateral side to the hip area

The edema failed to improve It was believed that perhaps the suture had not been carried high enough In January 1951 two additional sutures were implanted from an area of normal tissue below the ribs down across Poupart s ligament to the mid thigh The edema increased slowly during the next 14 months She complained of tenderness in the groin upon bending at the waist

In April 1953 she was admitted to the hospital and the sutures were removed At this time she was given an intensive course of hyaluronidase injected daily into the edematous tissue While in bed the edema subsided somewhat but when up and about even with her elastic support it recurred and reached its previous size When last seen on November 14 1953 the edema was slowly increasing as shown in the illustration Note the uniform swelling up the entire leg This type of case is the most difficult to manage She was seen only after the edema was massive A sympathectomy had been performed As indicated before this has now been found to be of no value in aiding lymphedema Actually the surgical incision and dissection further impede and damage the lymph channels

Case 36 Lymphedema Praecox


Note pale swelling from left foot to thigh


Side view


Wearing of knee length elastic stocking as part of treatment

Miss N M (The New York Hospital #639768) a 23 year old office worker was first seen on October 10 1952 She had a history of a gradually enlarging left leg from toes to groin over a three year period During this time she had had several episodes of cellulitis

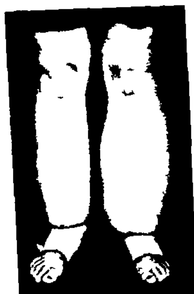
She was placed on the medical regimen No improvement was noted and one month later a suture was inserted extending from 5 cm above the inguinal ligament to 10 cm below it The following month she was given a series of three weekly injections of hyaluronidase The left figure shows a front view of both legs Note the pale swelling in the left foot to thigh The middle figure is a side view The right figure is a front view showing knee length elastic stocking being worn as part of her treatment (11) No dramatic improvement occurred

Case 37 Lymphedema Praecox



Mrs R W (The New York Hospital #292194) was seen in 1949 at the age of 42 See the pale swollen leg in the accompanying illustration She gave a history of swelling of the right ankle at the age of 18 The swelling gradually increased in size until the leg became cumbersome and heavy She was placed on the medical regimen and obtained some improvement The measurements decreased and the leg became less tense One month later she was admitted to The New York Hospital Under general anesthesia Dr Jere W Lord Jr inserted double strand No 0 nylon sutures laterally and medially from the ankle to the right lower quadrant of the abdomen Slight improvement was noted during the hospital stay probably due to the prolonged elevation Her subsequent course has shown no improvement

Case 38 Massive Lymphedema Praecox



This young woman noticed swelling of her right leg shortly after puberty The swelling gradually extended up both legs and slowly increased to its present massive size

The accompanying figure shows a front view of both legs The greatly swollen left leg hangs down over the ankle like a bag

Case 39 Lymphedema of Leg Secondary to Surgery in Groin



This 30 year old woman had a radical pelvic *operation with resection of lymph nodes* in her right groin for carcinoma of the cervix one and a half years ago Swelling of the right leg commenced shortly after operation and *slowly increased in size*

The illustration shows a front view of both legs Notice the pale edema of the entire right leg extending up to the groin

Case 40 Lymphedema of Hand

Case 40 Lymphedema of Hand



This 15 year old boy has lymphedema of the hand (see the illustration) When a baby he fell asleep with a rubber band around his wrist By the time his parents noted it some 12 hours had elapsed The hand was swollen It continued in a swollen condition throughout adolescence Treatment consisting of elevation and hydrotherapy has greatly improved the condition

Cases 41, 42, and 43 Stasis Ulcerations and Edema



Large ulcerations of ankles and edema of feet in a far advanced arthritic patient



Ulcerations and gangrenous areas in patient immobilized with arthritis



Gangrenous heels from pressure of lying in bed and immobility in patient with rheumatoid arthritis

Sitting with the legs in dependency for long periods of time results in poor venous and lymphatic drainage Even in normal individuals the legs may become swollen With inadequate return venous or lymphatic circulation this may at first be temporary After a period of months or years of relative inactivity and stasis the tissues break down and ulcerations develop Prevention consists in having these patients walk if possible Treatment should be directed at the underlying arterial or venous condition An oscillating bed is often helpful

The upper left figure shows stasis edema of the feet and large ulcers of the ankles The major arterial and venous systems are normal This patient was a far advanced arthritic and had not walked in many years

The upper right figure shows ulcerations and gangrenous areas in a patient immobilized with arthritis Old phlebitic lesions are present The major arterial supply is normal The

gangrenous lesions sloughed and healed when motion was instituted

The lower figure shows stasis eczema and gangrenous heels which developed from the pressure of lying in bed and immobility in a patient with rheumatoid arthritis

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4

Vasospasm and Diseases in Which It Plays a Major Role

When the body is chilled the peripheral arteries and arterioles are constricted and blood flow is reduced As time passes the oxygen content of the peripheral blood is lessened the lips ears nose hands and feet become blue the arms and legs turn a mottled purple If chilling is severe some areas may blanch Fever on the other hand results in marked peripheral vasodilatation All pulses become augmented The skin becomes hot pink, and dry

A slight change in temperature of a large surface of the body or a profound change in a small area can evoke a vasomotor response either constriction or dilatation This is mediated by reflex nervous action If blood returning to the vasomotor center in the brain is cooled peripheral vasoconstriction results The control of the flow of blood to the periphery is somewhat similar to the thermostat regulating water flow in the radiator of an automobile above a given temperature the water will be sent through the radiator (the peripheral circulation) in order to expel heat below this temperature the water is kept circulating in the interior of the motor (the deeper tissues) to conserve heat

The familiar blush that follows embarrassment is evidence that emotional factors influence peripheral blood vessels Anger and fear produce a cool pale moist skin

If the medium sized arteries are thickened from a disease as in scleroderma atherosclerosis obliterans or thromboangiitis obliterans the constriction that occurs following chilling may cause complete obstruction of the entire vessel A dead cadaverous white area develops along the distribution of the artery If the affected person comes into a warm room or increases his body heat for example by taking a hot drink the spasm may be released The blanched areas slowly change to blue as the blood trickles through the capillary bed and its oxygen is rapidly removed Soon a bright red flush appears as full dilatation of the vascular bed occurs (reflex hyperemia) and blood containing large amounts of oxygen floods into the venules which influence in large measure the color of the skin

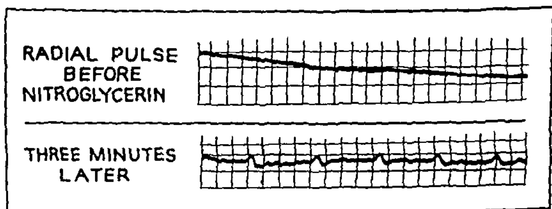
Disorders of these mechanisms lead to several clinical syndromes If periodic blanching of fingers toes ears and sometimes the nose occurs it is called *Raynaud's phenomena* or *syndrome* If blueness and coldness of the fingers toes hands or feet occur for long periods of time then the condition is termed *acrocyanosis* If the most prominent feature is mottling of the arms and legs with episodes of coldness and blueness as well it is called *livedo reticularis* (Dermatologists call it cutis marmorata) These conditions may be primary or they may develop secondarily to almost any of the dozens of vascular diseases

PRIMARY RAYNAUD'S DISEASE

This is a periodic blanching of digits, ears, and nose as explained above. It is precipitated by chilling or emotional stress. It often starts in childhood and lasts until middle life. It is far more common in women.

The abnormality is probably a function of the blood vessels themselves. The arterioles are often the site of the constriction. Severance of sympathetic nerves to the vessel results in a temporary improvement but later the vessels usually regain their sensitivity to cold and emotion even though denervated, *indicating that it is the vessel itself which is abnormal.* Hence sympathectomy usually provides only temporary relief.

These patients should dress unusually warmly. They should wear boots and gloves even on slightly chilly days. The application of heat directly to the blanched part should *be avoided. This may precipitate gangrene* as in Case 44. A spasm of the large and intermediate sized vessels, for example the radial, ulnar and digital arteries, can usually be quickly released by warming the entire body. Coming in out of the cold to a warm room or drinking a hot beverage are suitable measures. Nitroglycerin under the tongue acts quickly and effectively *to release the spasm* of the larger vessels but has less effect on the arterioles.



Good effect of nitroglycerin upon pulsations of radial artery in a 28 year old patient with vasospasm associated with Raynaud's disease. Impalpable before nitroglycerin, the pulsations became readily palpable after the drug.*

* Denicotinized cigarettes impregnated with nitroglycerin dilate the peripheral arteries equally as well as sublingual administration.

Case 44 Raynaud s Disease



This young woman had had cold moist hands and feet since childhood On slight ex posure to cold her fingers would turn a ca daverous white When she became warmer the fingers would gradually turn blue and then pink This change in color was blotchy Some areas of a finger would be white while an other area was turning blue and still other parts were becoming pink

At the age of 28 during a winter she sud denly experienced more frequent and longer lasting attacks in her left second finger Dur ing one such attack she soaked the finger in hot water The need of the tissues for oxygen became greatly increased Blood flow was insufficient to furnish it death of the tissues ensued

The accompanying figure shows a close up of this finger which is necrotic

She was treated by inducing fever through intravenous injections of typhoid vaccine Complete healing was obtained in three weeks time She was advised to protect her self against chilling There have been no fur ther episodes of gangrene

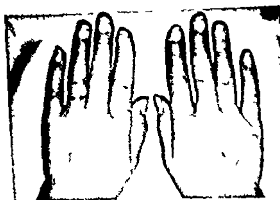
ACROCYANOSIS

This is a condition usually found in young women The hands and often the feet are persistently cool moist and blue during the winter Cold sweat may be profuse In summer there may be some improvement The skin shows little change except in color and temperature Scleroderma is never present The characteristic intermittent blanching and ruboric phases of Raynaud s disease do not occur Paresthesia is a frequent complaint

Experimental studies of Lewis suggest that it is probably the arterioles of the skin that are affected The mechanism of this disease rests in an unusual sensitivity of these vessels to cold The larger and medium sized vessels are patent

Many of these patients are mentally or emotionally disturbed persons The condition is not serious Gangrenous ulcers do not form except from unhygienic practices or overheating Treatment consists in keeping the body warm Vasodilating drugs have been disappointing Sympathectomy gives temporary relief only

Case 45 Acrocyanosis



Mrs J S (The New York Hospital #592 606) was 51 years of age For 10 years this middle aged farm woman had complained of episodes of severe pain and numbness in her hands and feet At such times they became a deep purple in color and were cold and wet Since childhood her extremities had been notably cool and moist All peripheral pulses were feeble

Sublingual nitroglycerin gave relief for several hours at a time Table 5 gives the oscillometric readings in her foot

The illustration shows the cyanosis and lack of scleroderma

Table 5 Effect of nitroglycerin on pulses of patient with vasospasm and acrocyanosis

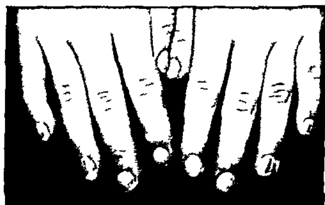
	2 to 5 min			
	Before Drug		After Drug	
	Right	Left	Right	Left
Blood pressure	128/78		118/70	
Pulse	68		72	
Radial pulse	2+	2+	3+	3+
Ulnar pulse	0	0	+	+
Femoral pulse	2+	2+	3+	3+
Dorsalis pedis	0	0	3+	3+
Posterior tibial	0	0	3+	3+
Oscillometric foot	0	0	0 5	0.5
Oscillometric above ankle	2 0	2 0	4 0	4 0

LIVEDO RETICULARIS

Adolescent girls and young women sometimes exhibit a mottled blueness of the arms and legs especially when they are slightly chilled This represents a constriction of a few arterioles often associated with a flaccid dilatation of the subcutaneous venules In long standing cases a nonspecific vasculitis is often found by biopsy This is usually accompanied by increased vasomotor tone in the hands and feet The peripheral pulses become faint, the hands and feet are cold moist and mottled but pale

This condition is usually benign but often excites a great concern on the part of parents It may persist for a decade or even two but usually decreases in severity thereafter Rarely small painful ulcers may occur Sympathectomy is of little value and is unwarranted Patients should dress warmly and avoid chilling Vasodilating drugs are helpful but their side effects are usually worse than the condition itself

Case 46 Livedo Reticularis



This 20 year old girl had increased vasomotor tone since early childhood Her hands and feet were always pale cool and moist except when the vasospasm was relieved by warmth fever or drugs Pulses were faint On exposure to a breeze or a cool atmosphere a mottled purplish discoloration developed on her arms and legs

The illustration shows a close up of the hands Note the blotchy discoloration

Case 47 Livedo Reticularis





In this patient ulceration developed This 30 year old housewife had had blotchy purplish discoloration of her arms and legs since childhood It was accompanied by greatly increased vasomotor tone as manifested by cool pale moist hands and feet and faint pulses

Ulcerations began to appear 11 years ago They were painful In the illustration note the cyanosis and black gangrenous areas at finger tips They recurred in cold weather and healed up in warm weather This suggested a frequently confused condition known as pernio or chronic chilblains Healing was obtained by artificial fever therapy The increased vasomotor tone was successfully combated by the strict avoidance of even slight chilling

FROSTBITE, PERNIO, CHILBLAINS

Overexposure to cold results in severe vasoconstriction If the exposure is such as to freeze the tissues serious damage to the vessels may occur This damage which may persist for years manifests itself by increased vasomotor tone which in turn may produce dermatitis intense itching and burning profuse perspiration with cold clammy hands and feet and occasionally ulcerations

Case 48 Frostbite (Chilblains, Pernio)



This middle aged woman had suffered frostbite of her legs 15 years previously Each winter after this she suffered pain numbness and discoloration of her legs She showed a very markedly increased vasomotor tone the feet were cold pale to purplish and moist at all times Neither the dorsalis pedis nor the posterior tibial pulses were palpable During the summertime she was reasonably comfortable but as soon as the cold weather began she had great discomfort throughout the entire winter except when she stayed indoors and kept herself warm On examination it was found that the pulses became bounding after the sublingual administration of glyceryl trinitrate indicating release from a severe degree of arterial spasm

The figure which is a photograph of both legs shows the intense redness ascending to the knees where it became streaky Despite the immediate response to nitroglycerin this patient did not respond therapeutically to any of the vasodilating drugs A bilateral lumbar sympathectomy was performed and an excellent response obtained The feet became warm dry and pink and her discomfort disappeared A five year follow up finds her in continued good health When the ulcers of pernio are developed the long term effects of sympathectomy are not usually satisfactory

Case 49 Frostbite with Ulceration

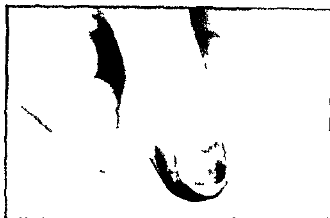


This 28 year old telephone operator developed frostbite of her hands six years previously All of the fingers healed except for the tip of the left index finger which became necrotic and open ulcers appeared

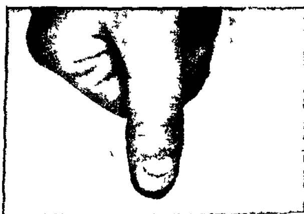
The illustration is a view of the hand showing the necrotic finger tip

In this patient healing was obtained by fever therapy The patient was given a course of induced fever therapy using intravenous typhoid vaccine The first injection was 5 000 000 bacteria A slight chill was obtained with a rise in temperature to 101° F The elevated fever continued for three days A second injection was given Again a fever was obtained for several days A total of four injections in a two week period resulted in healing

Case 50 Frostbite



Ten days after freezing



Four months later

This 28 year old registered nurse went skiing in January 1957 in Canada in frigid weather She came in for lunch and noticed that her hands were red and cold Her left thumb tingled and hurt

She skied again in the afternoon returning at 4 30 p m The thumb was white By nightfall it had turned dark purple

The treatment consisted of a petroleum jelly dressing and the administration of oral priscoline Hot soaks and heat to the thumb were scrupulously avoided

The left figure was taken 10 days after the incident Note the purple area under the nail and in the soft tissues

The right figure was taken four months later The dead skin areas have been replaced Nail regrowth is still incomplete

Case 51 Frostbite associated with Neurovascular Syndrome of the Shoulder Girdle

This 45 year old outdoor laborer developed frostbite with gangrene of the tips of fingers 2 3 and 5 Examination revealed a significant change in the strength of his radial pulse in various positions of the arm In a position of moderate hyperabduction the pulse vanished It was dampened by the costoclavicular maneuver and by turning the head sharply

Patients whose blood flow is reduced in various positions of the arm are apt to develop frostbite on mild exposure to cold

SCLERODERMA

This is a chronic disease of collagenous tissue that *may remain localized to the skin for years or may involve every organ* For many years its chief signs may be those associated with vasospasm The first sign of the disease may be typical Raynaud s phenomenon

Gradually the skin of the fingers thickens It becomes taut shiny glazed white and cool Small painful trophic ulcers occur at the finger tips which heal slowly leaving small dimples in the skin The tissues contract producing flexion deformities Later the skin of the arms legs feet face and torso thickens In far advanced cases even the breast abdomen and thighs are affected The lips become thin and drawn The mouth cannot be opened widely

Slight exposure to sun produces widespread dense deposits of melanin on exposed surfaces which do not fade Telangiectasis develops on the face hands anterior chest and elsewhere The bones in the terminal tufts of the fingers tend to become absorbed Entire phalanges and metacarpal or metatarsal bones can disappear

Calcium is deposited in the bursae about *the joints in tendons and sometimes in tissue* spaces These may ulcerate and protrude through the skin

Difficulty in swallowing is frequently experienced This is due to a thickening of the *esophageal* wall A diagnostic pattern is seen on x ray examination of the esophagus with barium The small intestine is sometimes in*volved resulting in diarrhea and obstruction* We have one patient who had 10 feet of intestine resected Scleroderma was not suspected at the time and the pathologist had not been able to establish a satisfactory diagnosis of the specimen One year later when we saw her she had developed the typical tight skin of the face and hands A review of the pathologic slides was found to be consistent with the diagnosis of scleroderma

A diffuse fibrosis of the lungs may occur Mr T (see the lower figure for Case 56) had had scleroderma for some 10 years Sympathectomies had been done to no avail This procedure is useless in this disease although it is *still* being done in some areas He flew to New York for consultation The slightly rarified air of the airplane cabin pressurized to 8 000 feet was sufficient to produce acute oxygen lack and great distress

These patients gradually resemble one another and appear as if they were siblings

TREATMENT

There is no specific therapy The disease may become stationary at any stage and is subject to remissions and exacerbations Whatever drug is being used just prior to a remission may get the credit for the improvement In the past three decades reports of success *with treatments have been published Critical* analysis however has failed to confirm the value of any Testosterone ACTH cortisone AT_{10} vitamin D PAS and a host of vasodilating drugs have been tried Acute phases of the *disease such as intestinal obstruction seem* to respond to steroid hormones at least temporarily

The precautions against chilling (outlined above under Raynaud s disease) should be carefully observed With the advent of cold weather very painful ulcers form on the fingers and are difficult to heal Such patients are *frequently helped by living in the tropics* —at least during the six months from November to April

As stated previously sympathectomy is of no avail While it may cause temporary warming of the hands the vessels soon regain their susceptibility to cold Ulcers form and the course of the disease is unaffected

Case 52 Advanced Scleroderma



Note tight shiny skin and deposits of melanin



Mouth open to full extent Note tight thin lips taut skin over cheek bones and great limitation in opening the lips

This 30 year old woman had developed spasmodic blanching of her fingers five years before Two years ago the skin of her hands became tight and shiny

The left figure shows the tight shiny skin The motion of the fingers was greatly limited by the skin contractions and became fixed in a clawlike position There was also discoloration by deposits of melanin

The right figure shows the mouth open to its full extent Note the tight thin lips the taut skin over the cheek bones and the great limitation in opening the lips This often produces dental problems and in rare cases the angles of the mouth have had to be split to permit extraction

Case 53 Advanced Scleroderma

This 40 year old housewife has had symptoms in her hands for more than seven years The disease started first with blanching of the fingers upon exposure to cold She noted that when she went to the icebox to take out a bottle of milk or if she went out of doors unless thoroughly protected against cold the fingers turned white In the summertime if she bathed in chilly water the fingers blanched When she came in out of the cold or took a hot drink the whiteness gave way to blueness and in turn to bright red discoloration As the years passed the skin of the fingers and toes became shiny and tight Later this tight skin became apparent also on the arms and on the face Each summer when her skin tanned the pigmentation remained through the fall and winter fading very slowly

The upper left figure is a close up of the face This shows marked tightness and shininess of the skin The lips are very thin red spider like hemangiomas are present These are quite characteristic of the late stages of scleroderma

The upper middle figure is a side view Notice how shiny the skin is and how taut it is over the bones of the arm

The upper right figure is a posterior view Note the tight skin over the scapulae which flare out from the body This posture is characteristic in advanced cases

The lower figure shows the hands The five characteristic points of scleroderma are well illustrated here 1 the clawlike appearance 2 the lack of motion in the joints—this is not due to any arthrodesis of the joints but is rather due to a constriction of the skin and soft tissues about the joints 3 there are some areas of ulceration and scarring over certain of the phalangeal joints notably over the left second 4 areas of pigmentation and depigmentation are seen at the base of the right second and first fingers 5 characteristic clubbing which follows prolonged atrophy as seen in the tip of the right third finger X ray study shows absorption of the tufts of the fingers and the deposition of calcium in subcutaneous areas and in tendons in bursal zones

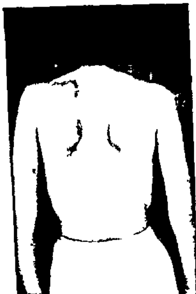
Note tight shiny skin thin lips and red spider like hemangiomas

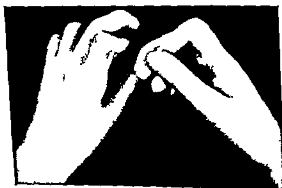


Note tight shiny skin over the arm bones



Note flaring of the scapulae





Note tight shiny skin clawlike appearance areas of pigmentation and depigmentation immobility of joints areas of ulceration and scarring

Case 54 Early Scleroderma



This 58 year old housewife noticed six months ago that the skin of her hands and arms had become much tighter and harder and more shiny than formerly Exposure to cold produces blanching and purplish discoloration of the fingers and hands There is increased pigmentation in areas exposed to sunlight This case is quite characteristic of the disease in the early stage (see the illustration) when the diagnosis is frequently missed Such cases frequently start as Raynaud s disease and years may pass before the scleroderma becomes obvious An early fluoroscopic or x ray examination of the esophagus with a barium swallow may show the characteristic stiffening of the rugal folds and peristaltic waves

Case 55 Advanced Scleroderma in a Man





This patient is a 33 year old salesman from Texas This is of interest because of the moderate to hot climate Fifteen years previously at age 18 he first developed episodes of Raynaud s syndrome By the age of 22 it was noticed that the skin of his fingers and hands was quite thickened A bilateral cervical thoracic sympathectomy was performed No benefit was obtained and a second bilateral cervical sympathectomy was done This too failed to produce any improvement This is quite characteristic of the effect of sympathectomy in this disease There may occasionally be warming of the skin for several months but the underlying disease progresses inexorably without any effect from the sympathectomy

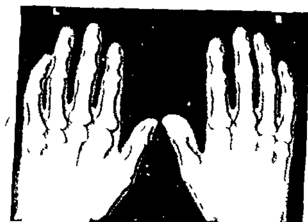
The left figure shows the advanced scleroderma in the face and chest

The right figure is a close up of the hands Notice the tight shiny skin and the clawlike position of the fingers

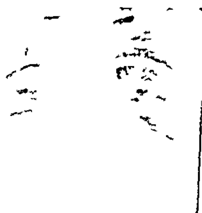
Case 56 Far Advanced Scleroderma with Pulmonary Changes



Extension of hands limited by tight skin



Radiograph of hands showing osteoporosis and absorption of tufts



Radiograph of chest showing extensive pulmonary fibrosis in both lungs

This middle aged man had noticed tightness of his skin two years previously In addition he had developed spasmodic blanching of the fingers and toes upon exposure to cold As the months progressed he noticed increasing shortness of breath He decided to fly to New York for consultation En route in the reduced oxygen tension of the high altitude he became acutely ill from oxygen lack and almost expired on the plane Upon admission to the hospital his vital capacity was found to be greatly reduced

The upper left figure shows the hands to be white tight and shiny They are extended to the fullest extent possible Note the limitation of motion of the joints This limitation is due to the tight skin

The upper right figure is an x ray of the hands showing osteoporosis Note the complete absorption of the distal two thirds of the terminal phalanx of the right thumb This is quite characteristic of scleroderma

The lower figure is an x ray of the chest and shows extensive pulmonary fibrosis in both lungs

Case 57 Early Scleroderma



Characteristic telangiectasia and pigmentation of face



Tight shiny skin of hands

This 40 year old secretary had had Raynaud's phenomena for one year Examination disclosed sclerodermatous changes in her hands arms face chest and legs

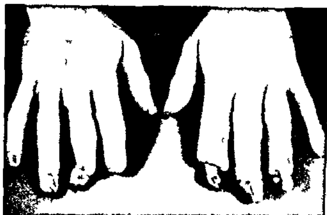
The left illustration is a view of the face and demonstrates the telangiectasia and pigmentation which are characteristic features of this disease Her mouth is opened to its fullest extent but is not as limited as in the other cases of further advanced disease

The right illustration is a close up of the hands showing the tight shiny skin The joints are stiff but no contractures have yet developed

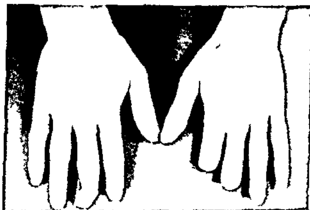
THROMBOANGIITIS OBLITERANS

The development of spasmodic blanching of the fingers is sometimes the earliest sign of this disease When observed in an adult male below middle age a careful search should be made for the other stigmas of the disease e g a blocked ulnar radial dorsalis pedis or posterior tibial artery or a history of recurrent phlebitis in a smoker

Case 58 Thromboangiitis Obliterans



Gangrenous hands before therapy



Healing after therapy

This is the history of a physician in early middle age Before we first saw him he had had three attacks of superficial phlebitis He then developed spasmodic blanching of the fingers and toes after slight chilling of the body

Ulcerations developed on the finger tips Examination showed sluggish radials and occluded ulnars He was hospitalized He had been accustomed to smoking 20 cigarettes daily but stopped this habit completely during the first hospital day He was given fever therapy in the form of intravenous typhoid vaccine Using a dilution of 100 million organisms per ml five million bacteria were given the first day Four hours later he had a slight chill followed by a fever of 101° F We prefer to obtain two to three degrees of fever without a chill but this is not always possible The temperature remained elevated for two days On repetition of the treatment no fever was produced The dose was therefore increased by three million organisms every fourth day Healing was well advanced in three weeks at which time he went home He returned to a busy general practice For some six months he succeeded in avoiding tobacco However during a period of great stress he resumed smoking The disease promptly became active and new ulcers appeared

The left figure shows the gangrenous hands The radial ulnar dorsalis pedis and posterior tibial vessels were occluded Because of the intense pain he had become addicted to narcotics

He was again admitted to the hospital With great difficulty he gave up tobacco Narcotics were reduced then successfully omitted Again fever therapy was given Collateral flow developed and good healing was obtained with only minor loss of tissue as shown in the right figure

If he should resume the use of tobacco the disease may be expected to involve other vessels such as the brachial iliac coronary cerebral or mesenteric arteries

ATHEROSCLEROSIS OBLITERANS

The digital vessels may be narrowed because of atheromatous deposits Slight chilling of the body produces constriction of those of the affected vessels that are still elastic and of the collateral arteries Because of the encroachment on the lumen of the atherosclerotic plaques the constricted vessel may become totally occluded Spasmodic blanching results

NEUROVASCULAR SYNDROMES OF THE SHOULDER GIRDLE

The large vessels and nerves pass from the thorax to the arm In so doing they traverse narrow areas between bones muscles and tendons Slight abnormalities in structure or in dynamics can lead to pressure or stretching of the vessels or nerves Episodes of vasospasm and paresthesia may result Even thrombosis of the subclavian vein or artery can be produced

A *cervical rib* even rudimentary may encroach on the vessels or nerves in certain positions

COSTOCLAVICULAR SYNDROME

Relaxation of scalenus and other neck muscles may result in pinching the neurovascular structures between the clavicle and the first rib

SCALENUS ANTICUS SYNDROME

The scalenus anticus may be hypertrophied or in an abnormal position It may compress the nerves and blood vessels in various positions of the head and shoulders

HYPERABDUCTION SYNDROME

This may occur in sleep as in Case 59 or in certain occupations such as ceiling painting or plastering

Case 59 Hyperabduction Syndrome

This 52 year old clerk developed blueness numbness and tingling of the right second third and fifth fingers The fingers were cold The responses to light touch and pin prick were diminished (upper left figure)

With the hands at the sides his radial and ulnar pulses were palpable When the hands were raised over his head the pulses disappeared It developed that he had the habit of sleeping with his hands in back of his head He was instructed to sleep with his hands at his sides Prompt and marked improvement resulted (upper right figure)

The lower left figure shows an anatomic dissection of a fresh cadaver before contraction of muscles has set in It shows the relation of the axillary artery and brachial plexus to the arms in a position of relaxed abduction Nerves and vessels are under no tension or pressure

In the lower right figure the arm has been placed in hyperabduction Note how the vessels are stretched around the coracoid process and pinched between the clavicle and first rib

Cyanotic pregangrenous fingers



Fingers pink following therapy





Arm in position of relaxed abduction



Arm placed in hyperabduction

Case 59 Hyperabduction Syndrome

This 52 year old clerk developed blueness numbness and tingling of the right second third and fifth fingers The fingers were cold The responses to light touch and pin prick were diminished (upper left figure)

With the hands at the sides his radial and ulnar pulses were palpable When the hands were raised over his head the pulses dis appeared It developed that he had the habit of sleeping with his hands in back of his head He was instructed to sleep with his hands at his sides Prompt and marked improvement resulted (upper right figure)

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Affected hand puffy and cyanotic



Arm swollen skin shiny and blue



Foley elastic sleeve in place

EFFORT SYNDROME

The axillary vein may be damaged and may thrombose from sudden repeated forceful muscular contractions as in pitching hay This has been called the effort syndrome It is due to trauma on the axillary vein by various structures of the shoulder girdle

AXILLARY VEIN THROMBOSIS

Another example of a shoulder girdle syndrome is illustrated by Case 60

Case 60 Axillary Vein Thrombosis

This 42 year old laborer fell and struck his right shoulder Following this the arm became swollen painful and cyanotic Whenever he attempted to use it motion was quite painful Examination disclosed normal pulses and normal oscillometric readings The circumference of the arm forearm wrist and hand was increased With the arm in dependency mottled cyanosis became apparent The collateral veins over the anterior shoulder and pectoral area were markedly increased and dilated

The upper figure shows the hands The *affected hand is puffy and bluish The swelling* is indicated by the marked decrease in the wrinkles over the first phalangeal joints

The lower left figure shows slight swelling and cyanosis with the arm in dependency

The lower right figure shows the Foley elastic sleeve in place This sleeve is designed to exert compression on the tissues and to limit the edema

Treatment for this type of case consists in elevation of the arm The arm is suspended in traction in a comfortable position and with counterweights which just balance the weight of the arm so that the patient is able to move about the bed or if seated to move about in the chair without difficulty or discomfort This continued elevation results in a marked decrease in the venous pressure together with an increase in return lymphatic flow The edema is soon cleared However many months transpire before sufficient veins recanalize and adequate drainage is obtained Anticoagu*lant drugs help to prevent* further extension of the thrombus peripherally They also prevent the development or extension of proximal thrombi which may embolize To summarize treatment consists in 1 anticoagulant therapy 2 elevation of the affected limb 3 compression with elastic bandages or sleeves and 4 hydrotherapy with cool baths and whirlpool baths are helpful

Case 61 Phlebitis (with Gangrene Secondary to Vasospasm)





This middle aged woman was admitted to the hospital with the extensive gangrene noted in the left figure with a presumptive diagnosis of arterial embolism Careful examination disclosed that the primary lesion was an extensive phlebitis with local vasospasm

Lumbar ganglionic blocks relieved the vasospasm Anticoagulation therapy was instituted elevation and hot moist packs were applied The gangrenous tissues sloughed and healing ensued The right figure shows the areas with healing with only minor loss of tissue

THROMBOPHLEBITIS

Acute thrombophlebitis in the veins of the legs may cause an irritation of the nerves or the artery accompanying them This is particularly true in Hunter s canal where the vessels are closely aligned Severe arterial spasm may result Usually this is relieved by warm moist packs Occasionally more radical measures are needed as in the following cases

PHLEBITIS WITH GANGRENE SECONDARY TO VASOSPASM

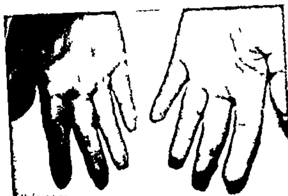
As mentioned above phlebitis in the legs especially in the femoral veins may give rise to a severe degree of vasospasm At times this may be so severe that superficial gangrenous areas develop especially in the toes

Sometimes it may be difficult to determine whether one is dealing primarily with an arterial disease or whether phlebitis is the primary disease The pulses in the feet and the oscillometric readings may be absent To release arterial spasm the simplest measure is to give nitroglycerin sublingually the usual dose is 0 0004 gm Ganglionic blocks are sometimes used This procedure has the disadvantage that it should not be used if the patient is on anticoagulant therapy because of the danger of deep seated hemorrhage from needle trauma The risk is slight but some serious hemorrhages have been reported Slight degrees of vasospasm can be relieved by warm moist packs These packs also help to clear the lymphatic channels

Case 63 Raynauds Syndrome with Gangrene Secondary to Arsenical Medication



Note necrotic gangrenous ulcerations on fingers



Following cervical sympathectomy hands became pink and warm

This 38 year old unmarried woman had had bronchial asthma for many years She had been given the usual treatment for asthma by her physicians and at times she was free of it but at other times she had severe exacerbations She heard of a supposed cure for asthma in the form of a secret medicine being dispensed by a physician in a distant state She traveled there and obtained the medicine While she was taking it her asthma greatly improved Accordingly she obtained a number of bottles and returned home She continued to take this preparation She soon began to develop periodic severe blanching of the fingers associated with numbness and tingling These episodes of Raynauds syndrome occurred at first only on chilling or during periods of emotional tension Later however they occurred frequently throughout the day without relationship to cold The numbness and tingling increased pain became very marked vasospasm increased thromboses developed in the digital arteries Necrotic ulcerations of the fingers developed She was admitted to the hospital

The left figure is a view of both hands Note the necrotic gangrenous ulcerations on several fingers

The right figure is a view of both hands following cervical sympathectomy Some improvement in release of the vasospasm occurred The black necrotic areas have localized

An analysis of this secret medicine proved it to be Fowlers solution which contains arsenic The disease in question was arsenic poisoning with severe peripheral neuritis secondary vasospasm and thromboses of the medium sized vessels

Case 62 Gangrene of the Toes Secondary to Phlebitis with Vasospasm



As in Case 61 this 46 year old housewife when first seen had gangrenous areas of her toes It was very difficult to determine whether or not the disease was primarily an arterial one Sublingual administration of nitroglycerin resulted in a return of the pulses and oscillometric readings She was treated with anticoagulant therapy elevation of the feet and heat applied to the abdomen She made an uneventful recovery without any major loss of tissue The black necrotic areas of the toes proved to be a gangrene only of the skin When this sloughed good healing was obtained

As seen in the illustration the dorsal aspect of the toes shows gangrenous blebs on toes 1 2 3 and 4

PERIPHERAL NEURITIS

This condition may be due to exogenous toxins such as alcohol heavy metals such as arsenic lead mercury copper gold silver drugs such as emetine isoniazid and chemicals such as nitrobenzol It may be due to deficiency states such as beri beri or pellagra The toxemias of infectious diseases such as diphtheria often cause a polyneuritis Periarteritis nodosa (polyangiitis) is a common cause While paresthesia numbness and muscular paralysis may be the most prominent features vasomotor changes are very *common In some such as Case 63 they are* the major factors

Case 64 (continued)

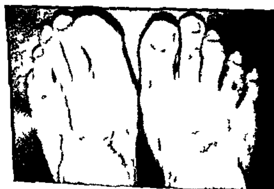
tomy was performed immediately The patient was placed on an oscillating bed reflex heat was used across the abdomen in the form of a heating pad one half hour on one half hour off Convalescence was slow but steady She was discharged one month after admission having recovered her major circulation and having lost only a small amount of tissue in the toes All pulses returned The accompanying photographs were taken during the recovery stage one week after admission They show spots of necrotic skin Earlier in the course of the disease the toes were cyanotic Two weeks after these pictures the feet were completely healed

The left figure shows the dorsum of the foot and toes Purplish areas of necrosis are present over the dorsum of the foot and toes

The right figure is a view of the plantar surface A large purple area is present over the metatarsal joints and the large toe

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Case 65 Ergotamine Poisoning



This middle aged building superintendent had suffered from migraine for many years He had polycythemia He had taken ergotamine tartrate at frequent intervals This had affected his headaches favorably For the two weeks prior to admission he had taken unusually large doses Gangrene developed in the right fourth toe

The accompanying photograph was taken It is a close up of the toes and shows a black area of gangrene surrounded by red pregangrenous tissue The foot was cold the vasospasm was severe

Treatment consisted of withdrawal of the ergotamine reflex heat applied to the groin the use of an oscillating bed and walking for increasing distances hourly during the day time Healing took place without difficulty

ERGOT POISONING

Medical history is replete with epidemics of ergot poisoning caused by the ingestion of rye bread made with grain contaminated by the fungus ergot This was common in medieval middle eastern Europe but several years ago such an outbreak occurred in a small town in France

Lewis performed a series of experiments using ergot derivatives In man single experimental doses (0 2 to 0 5 mg of ergotamine or 0 2 mg of ergotoxine) given intravenously caused giddiness pressure in the head and tiredness The skin of the face hands and feet became cyanotic for as long as 48 hours and profound rises in blood pressure occurred Experiments with roosters have shown that by feeding with infected grain or by daily intramuscular injections of ergotoxine (0 1 mg) complete stagnation of blood in the comb can be easily produced and maintained the result of continuous arterial spasm The stagnation leads to stasis The vessels are damaged and thrombosis follows

Ergotism is rare today except following the misuse of medications as in the case histories below

Case 64 Aute Ergot Poisoning with Gangrene of the Toes

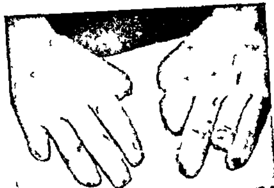




This 39 year old woman thought that she was pregnant She attempted to produce an abortion by taking fluid extract of ergot She became confused about the dosage instead of taking four drops at a time she took four teaspoonsful In a period of 36 hours she consumed 24 ml Both legs became extremely cold painful and later numb On admission to the hospital she was pulseless from the waist down Oscillometric readings were zero up to the groins Early gangrene had developed in the toes of the left foot A lumbar sympathec-

Case 66 Cryoglobulinemia

A



B





C

A B C Loss of terminal phalanges (Brown pigment is result of potassium permanganate soaks not to be confused with lesions)



D

D Loss of tip of nose and ear lobes

CRYOGLOBULINEMIA

In certain disease states abnormal proteins are formed which precipitate when cooled slightly below body temperature These precipitations occur most commonly in the acral parts where the temperature is lowest namely the tips of the fingers toes ears and nose The precipitate plugs small vessels and leads to necrosis of the tissues supplied These cold precipitable proteins have been found in patients with multiple myeloma lymphomas of all types and various leukemias

Case 66 Cryoglobulinemia

This 50 year old white male railway clerk had developed blanching of the fingers toes nose and ears some four years prior to admission to the hospital Gangrenous areas had developed on the tips of these areas and he had lost tissue on all the surfaces Study in the hospital showed him to have multiple myeloma Two to four grams of cold precipitable spontaneously crystallizing protein were found in his blood This case was reported by Barr Reader and Wheeler

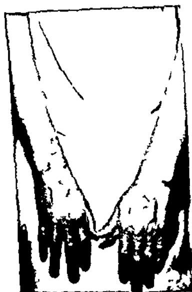
The upper left figure is a photograph of the dorsum of the hands showing the ulcerating amputation stumps of all digits

The upper right figure is a view of the palmar surfaces of the hands The brown pigment is the result of soaks with potassium permanganate and should not be confused with the lesions

The lower left figure is a view of the patient s feet showing ulcerating amputation stumps of toes

The lower right figure is a view of the patient s head showing a healed ulcer on the tip of the nose and scalloped edges of the ears secondary to gangrene

Case 67 Sudeks Atrophy (Traumatic Reflex Vasospastic Dystrophy with Osteoporosis following Fracture and Axillary Vein Thrombosis)



This elderly ships captain fell on the bridge of his ship during a storm He suffered a fracture of the right humerus The arm was placed in a cast Axillary vein thrombosis resulted This was unrecognized for several weeks The arm became edematous and painful he did not exercise it the muscles became swollen and taut Vasospasm was severe Radial and ulnar pulses could not be obtained unless the patient was warmed or sublingual nitroglycerin was given As time went on the fingers of the hand became thickened and immobile X ray showed marked osteoporosis Treatment consisted of 1 cortisone to combat the causalgic pain 2 reflex heat 3 physiotherapy in the form of gentle massage and warm whirlpool baths 4 active exercise Improvement was rapid Anticoagulants were also administered to prevent extension of the thrombus they were maintained until he was back to full duty

The photograph shows a view of both hands Note the swollen shiny right hand fingers and arm

Closely related to this disorder are crushing or tearing injuries to the hand In such cases the vasomotor disturbance leads to a cold hand rather than a warm one Raynaud s phenomena are common complications

CAUSALGIA, SUDEK'S ATROPHY

The most frequent cause of this malady is an injury to a peripheral nerve without dividing it The median or sciatic nerves are commonly involved but it may be the small nerve endings at the tip of a digit

Pain may develop within a week of injury and gradually increase in intensity It often results from the use of a cast for a minor or chip fracture It is burning in character and associated with extreme tenderness of the area of distribution of the affected nerve The pain is superficial and is elicited by slight contact by friction by warmth or by cold It may be so easily provoked and so severe that the patient guards his arm or leg closely The pain is intractable and lasts for many months or may even be permanent if not treated Cortisone may provide some relief

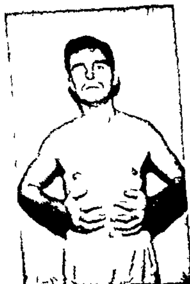
The skin may be pink and warm or cold cyanotic, and wet with sweat It becomes shiny Fingers lose their wrinkles The nails frequently stop growing and become brittle X ray studies show progressive osteoporosis

Treatment consists of increased activity physiotherapy massage weight bearing This must be pursued even if increased pain results temporarily If the reluctance of the patient can be overcome the results are usually good

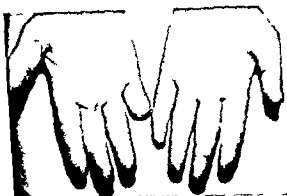
RHEUMATOID ARTHRITIS AND COLLAGEN DISEASES

All of the collagen diseases and related pathologic states (periarteritis nodosa dermatomyositis scleroderma lupus erythematosus rheumatic fever) may damage the peripheral vessels Raynaud s phenomena occur commonly in these conditions

Case 69 Raynaud s Syndrome Secondary to Rheumatoid Arthritis



Note spindle shaped pale sclerodactylic fingers



Note tense shiny skin and cyanotic fingers

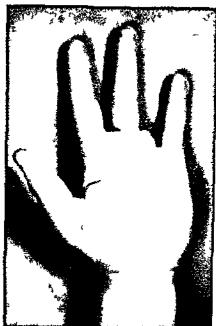
This 37 year old coal miner had suffered many injuries to his hands He developed painful white cold fingers The skin of the digits became thick and taut the joints became spindle shaped and the right ulnar artery was found to be occluded Eosinophilia was persistent and ranged from 6 to 11 per cent A very thorough study in the hospital led to the establishment of the diagnosis as rheumatoid arthritis

The left illustration is a front view of the patient with his hands placed on his abdomen Note the spindle shaped pale sclerodactylic fingers

The right illustration is a close up of the palmar surfaces of the hands Note the tense shiny skin The terminal No 5 right phalanx had been lost in an accident some years previously

Marked improvement in all symptoms was obtained with steroid therapy

Case 68 Persistent Vasospastic Phenomena following Trauma



Note mottling of skin with cyanosis of fingertips



Dorsal view

This U S Army private while in combat suffered a hand injury from a grenade The fourth finger was amputated When seen by us three years after the explosion the hand showed extreme vasospasm The palm of the hand is shown in the left photograph and the dorsum at the right The radial and ulnar pulses were very feeble The hand was cold moist and purple When nitroglycerin was administered sublingually the pulses became augmented indicating that the underlying mechanism was not an organic occlusive disease but rather on the basis of muscular contraction of the vessel walls The patient was instructed to avoid all factors that might induce vasospasm namely 1 tobacco 2 chilling of the body 3 chilling of the hand He was advised to wear a glove in any cold weather and he was instructed to take a warm drink when his fingers became blanched and if this did not give instant relief to take a nitroglycerin tablet under his tongue With the passage of time the symptoms gradually subsided

Both photographs show mottling of the *skin with cyanosis of the fingertips Of interest* is the fact that the brother of this patient was later shot in the hand and developed the same syndrome

SUMMARY

Vasospasm in the peripheral vessels has been discussed Primary diseases are Raynauds disease acrocyanosis and livedo reticularis Case histories were cited in which the clinical manifestations and therapy were discussed Secondary Raynauds phenomena may occur in the course of many diseases affecting blood vessels They include the following scleroderma thromboangiitis obliterans atherosclerosis obliterans neurovascular syndromes of the shoulder girdle thrombophlebitis axillary vein thrombosis frostbite pernio and chilblains peripheral neuritis ergot poisoning and ergotamine poisoning cryoglobulinemia causalgia Sudeks atrophy and traumatic vasospastic phenomena rheumatoid arthritis and periarteritis nodosa Cases illustrating these conditions have been presented

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PERIARTERITIS NODOSA

Vasospasm is frequently present in periarteritis nodosa it is often the presenting sign of the disease In the early stages it may be confused with *thromboangiitis obliterans* primary Raynaud's disease or Raynaud's phenomena secondary to other vascular diseases

Case 70 Periarteritis Nodosa



This patient is a middle aged salesman His illness *commenced two months previous* to the time these photographs were taken He noticed a sudden numbness in his left hand During the next several days two fingers became blanched and cold The other limbs rapidly *showed signs of arterial insufficiency* There was a leukocytosis with eosinophilia up to 10 per cent Peripheral neuritis developed probably due to ischemia of the nerves Thrombosis of the large vessels soon followed

The upper left photograph shows black gangrene of the toes with sloughing of tissues

The upper right photograph is a view of the plantar surfaces of the feet and shows extensive gangrene extending to the midportion of the left foot

The lower photograph is a view of the hands showing extreme pallor The left index finger is gangrenous and tissue is sloughed from the tip

5

Aneurysm

An arterial aneurysm in its primary form is simply an abnormal dilatation of an artery. Trauma may injure the vessel wall. The pressure of the circulating blood then causes the weakened wall to dilate. The dilatation may be diffuse or saccular. The sac consists of the vessels wall. This is a "true" aneurysm.

In other instances the arterial wall is split or destroyed in part or completely and the contiguous structures form a sac like enclosure through which the blood circulates. This is called a "false" aneurysm, which usually develops as a result of a fracture of a calcific plaque.

The inner wall of an aneurysm may be lined with laminated blood clots thereby causing a reduction of the pulsations.

Dissecting aneurysms are usually on the basis of median necrosis but may be of atherosclerotic origin. The original splitting of the media may occur without any communication with the lumen. At some time later a tear of the intima occurs. Dissection progresses between the layers in either direction. Less commonly a plaque splits and the medial dissection extends from there.

1 Congenital Aneurysms. In these the medial coat may be undeveloped or absent; dilatation may occur early in life. These aneurysms are often found intracranially in the internal carotid artery or in the circle of Willis. Rupture usually causes fatal cerebral hemorrhage although minor leakage may give advance warning.

2 Syphilitic Aneurysms. Aortitis, a manifestation of inadequately treated syphilis, occurs most often in the ascending aorta and the aortic arch, less often in the descending and the abdominal aorta, and more rarely *in the extremities*. These aneurysms may be fusiform or saccular.

3 Atherosclerotic Aneurysms. Aneurysm due to atherosclerosis is now encountered with increasing frequency. This is due primarily to the aging of the population. The medial coats of arteries affected by atherosclerosis may rupture. This may occur at an area of calcific deposit or plaque. The form of dilatation of the artery depends upon the local condition of the blood vessel, the contiguous structures, the tension in the vessel, and the degree of successful repair. Mild trauma of a vessel affected by atherosclerosis is a frequent precipitating cause of aneurysms. Common examples of this are saccular aneurysms occurring in the popliteal artery.

Atherosclerosis with medial necrosis is the most common cause of dissecting aneurysms. Thrombosis of the artery is a frequent complication and at times may produce complete occlusion. Rupture of an aneurysm of a large artery or in a critical location may result in death. Gangrene may result from aneurysm of a peripheral vessel. Usually this is associated with occlusive thrombosis of the aneurysm or with emboli which arise from a mother clot in the aneurysm and move peripherally to block smaller arteries. Early diagnostic cri

As the result of an arteriovenous fistula short circuiting occurs in which a portion of the blood returns to the heart without having accomplished its function of nourishing the tissues If these channels are of sufficient total size so that large amounts of blood may be shunted by this short circuit nutrition in the distal part may be inadequate for the life of the tissues Gangrene may ensue This may also result in cardiac hypertrophy and with large fistulas in heart failure In small fistulas this does not occur Arteriovenous anastomosis may be

1 Congenital Congenital anastomoses are much more common than has previously been believed The possibility of arteriovenous connections should be considered when there are congenital vascular anomalies such as cavernous hemangiomas suddenly appearing "varicose veins and some of the port wine and other vascular skin anomalies Some congenital arteriovenous connections are locally malignant in that after excision new anastomoses develop with extension of the process usually proximal Though pathologically they do not have the appearance of a malignancy they may ultimately result in the loss of the part by amputation and in some instances in the loss of life

2 **Traumatic** Gunshot or stab wounds are common causes of arteriovenous fistulas but they may be caused by crushing injuries as well

3 **Secondary to Malignancy** Malignancy may weaken the walls of a blood vessel by direct invasion resulting in an arteriovenous fistula

4 **Secondary to Bacterial Infection of the Surrounding Tissues** Such infection may invade the walls of an artery and vein producing an arteriovenous fistula

5 **Secondary to Fungous Infections** Such infections may act similarly

teria are the physical findings of a *pulsating* mass or the roentgenographic appearance of enlarged vessels often with calcified walls Pain may occur especially if there is erosion of bone or pressure on nerves Bruits can frequently be heard over the aneurysm but can also be heard over any narrowed or roughened artery

4 Mycotic Aneurysms The wall of the aorta or of other arteries may be weakened by suppurative processes secondary to actinomycosis tuberculosis subacute bacterial endocarditis septicemia pneumonia typhoid fever or other infectious diseases A local infection around a major vessel may so weaken it as to lead to an aneurysm or to rupture

Aneurysms occur most often where there is marked stress and strain on the arterial walls, e g near joints in the extremities

5 Traumatic Aneurysms May be of the true" or "false variety An injury to an artery may cause its immediate or later dilatation This development may be delayed due to the pressure of the contiguous structures or because of a concomitant hematoma

6 Embolic Aneurysms An embolus may enlarge an artery and cause a weakening of the intimal and medial layers with secondary aneurysmal dilatation

7 Idiopathic Aneurysms In certain instances the true cause for the aneurysm cannot be ascertained

ARTERIOVENOUS ANASTOMOSIS (FISTULA)

Arteriovenous anastomosis is an *abnormal* communication between an artery and a vein This may be congenital or acquired The communication may be

1 Direct such as follows a stab or gunshot wound
2 Through a saccular aneurysm involving primarily one vessel
3 By an abnormal dilatation of the walls of the several vessels involved
4 By arteries and veins opening into contiguous structures

Congenital anastomoses are usually multiple and may number in the thousands

The following signs and symptoms may indicate the presence of an arteriovenous anastomosis

1 Enlargement of a limb or area of the body surface
2 Local increase in perspiration and growth of hair
3 Increased local temperature
4 Rubor of the affected area
5 *Unusual prominence of veins*
6 An audible bruit over the affected area occasionally a thrill may be felt In congenital or small anastomoses bruits are frequently absent
7 Increased oxygenation of venous blood proximal to that area This causes the venous blood to be brighter red than the blood from corresponding normal veins Such a comparison constitutes a simple useful diagnostic test Studies for oxygen unsaturation usually confirm this finding
8 Cardiac insufficiency with enlargement of the heart is a frequent complication
9 The pulse may be slowed by pressure applied to the affected area
10 Arteriographic studies may show the abnormal communication
11 Birth marks hemangiomas and related vascular lesions may be associated

Case 71 Abdominal Aortic Aneurysm

In the accompanying figures note the contrast media in the aorta in the two films taken one second apart The colon is partly filled with barium

Accordingly he was prepared for operation The aneurysm was resected An aneurysm of the abdominal aorta below the renal arteries (such as in this case) is almost always arteriosclerotic Thoracic aneurysms were formerly most frequently syphilitic but as a result of the improvement in antisyphilitic therapy and the aging of the population this is no longer true This aneurysm was replaced by an aortic homograft extending from 3 cm below the renal arteries to each common iliac artery The patient withstood the procedure well Pulses were obtainable in both femoral arteries immediately following the operation and also in the right foot Pulses did not return to the left foot until the following day The wound healed well There was postoperative distention He was kept in bed for 14 days At the end of that time he was progressively mobilized and at the end of the twenty fourth day was discharged from the hospital He has been followed for three years He has regained his strength feels well and fit His pulses are strong in all normal locations

Without surgical intervention the prognosis of this patient was hopeless In a study of patients with abdominal aortic aneurysms seen at The New York Hospital it was found that fewer than 5 per cent survived a five year period if unoperated DeBakey has shown that following surgery the longevity is about the same as the normal for the age group Until the development of artery banks the problem of the abdominal aortic aneurysm was of academic interest only The successful use of the graft in this patient shows the possibilities of surgery in this condition which had formerly been considered hopeless Today dacron and nylon grafts have largely replaced homografts

Case 71 Abdominal Aortic Aneurysm




Contrast media in aneurysm of aorta colon partly filled with barium

Mr A B (The New York Hospital #744 164) was a 71 year old private chauffeur who had retired to live in Florida In June 1956 he developed dyspepsia He consulted a physician who prescribed alkalies His symptoms improved X rays were taken at that time which showed a mass pressing against the stomach This was thought to be an aortic aneurysm and he was referred for vascular consultation

General examination disclosed that the patient was a vigorous man The abnormal findings were largely confined to the abdomen At the level of the umbilicus there was a firm pulsatile mass measuring 10 cm in width and 11 cm in length it was slightly tender to pressure Over this mass a rough systolic bruit could be heard This bruit could be traced up into the chest and was synchronous with the systole of the heart Also over the area of the aortic valve there was a similar rough systolic bruit Both femoral pulses were strong as were the dorsalis pedis and posterior tibial pulses Oscillometric readings were normal blood pressure in the right arm was 172/96

Gastrointestinal series showed extrinsic pressure with displacement of the stomach duodenum and small bowel due to an enlarged calcified abdominal aortic aneurysm An aortogram was performed under general anesthesia This showed a large saccular type of aneurysm

There was no nausea vomiting or diarrhea There were no bloody or tarry stools He was admitted to the hospital via ambulance On admission to the hospital 15 mg of morphine was administered for the pain During the night, on two occasions he had some nausea and vomiting but slept for several hours

The blood pressure and pulse were well maintained Physical examination showed that he was a well developed and nourished acutely ill man who was apprehensive per spiring freely and in acute pain but alert and oriented No cyanosis or pallor was noted His pupils were round regular equal and reacted to light and accommodation Medial and just below the angle of the left scapula an area of dullness to percussion and bron chial breathing was noticed No rales were heard Retrosternal dullness in the upper sternal area was found but over all cardiac dullness was not increased A suggestive para doxical pulse was noted The second aortic sound was greater than the second pulmonic sound The rhythm of the heart was regular and the sounds were somewhat overshadowed by a loud to and fro friction rub heard over the whole pericardium The neck veins were not distended the liver and spleen were not felt and the peripheral pulses and femoral pulses were palpable and equal Neurologic examination was normal

During the night, in addition to the peri cardial friction rub a high pitched diastolic murmur was heard at the base of the heart Peripheral pulses were still palpable and of good quality No venous distention occurred Death occurred precipitously

Postmortem examination was conducted The illustration shows the extensive dissecting aneurysm that was found extending from the sinus of Valsalva rupturing into the peri cardium and dissecting the medial coat of the artery down through the chest into the abdominal aorta and ending at its bifurcation The pericardium contained about 700 ml of blood Extensive atherosclerosis of the ab dominal aorta was found together with ather omatous formations in the coronary vessels occlusion of the anterior circumflex coronary artery and a healed myocardial infarction in the anterior lateral wall

COMMENT In retrospect one would say that this man s basic disease was hypertension of unknown cause This led to extensive atherosclerosis of the aorta and coronary ar teries A thrombotic occlusion had developed at the site of an atherosclerotic plaque in a large coronary artery causing a myocardial infarction Medial degeneration had occurred in the aorta dissection from or into an athero sclerotic plaque had led to a splitting of the layers of the aorta in the region of the medial degeneration This in turn extended the dis section down the full length of the aorta A rupture into the pericardium from the original site of dissection had also occurred The dis section may have been progressing silently for a long time but extended with critical splitting occurring on the day before death when the patient experienced the severe pain in his chest

The differential diagnosis for consideration was a fresh myocardial infarction In such a case one would not expect the blood pressure to have been maintained The finding of car diac tamponade with a hemopericardium might have been ascribed to the anticoagulant therapy This however was not the case Anticoagulants were administered in an effort to prevent new thrombus formation in the diseased coronary tree This therapy does not appear to interrupt the atheromatous process which may eventually occlude the coro nary vessels completely in the absence of a thrombus

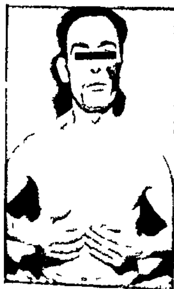
Case 72 Dissecting Aneurysm of the Aorta



Patient E A K The New York Hospital #740148 was a 54 year old actor The dissecting aneurysm of the aorta is shown in the accompanying figure In May 1953 he suffered an anterior lateral myocardial infarction

For one year prior to this he had been known to have had high blood pressure At the time of his infarction his blood pressure fell to normotensive levels He was in the hospital for a period of three weeks on anti coagulant therapy He was then ambulated His blood pressure gradually rose and within a period of six weeks returned to the pre infarction levels During the next two years the blood pressure ranged from a high of 190/110 to a low of 160/90 He was continued on long term dicumarol therapy His pro thrombin time was maintained between 24 and 32 seconds on an average daily dose of 75 mg of dicumarol During two years of observation he showed no untoward effects from the anticoagulant administration His blood cholesterol level in October 1955 was 250 mg per cent Following a diet low in animal fat it was found to be 192 mg per cent in March 1956 His serology was negative his hemogram was normal X rays of the gallbladder showed normal function Serpasil and Raudixin were used in an attempt to control his blood pressure However even small doses of these drugs gave rise to nasal stuffiness and a regular dose schedule was not followed

On May 30 1956 three days prior to his death his prothrombin time was 28 seconds with a control of 14 seconds On that day he had no complaints During the early morning hours of June 2 he was awakened by a severe bifrontal headache which was not relieved by aspirin He finally fell asleep and on arising remained well until shortly after lunch when a sudden severe nonradiating persistent substernal pain developed Weakness perspiration and marked apprehension with poorly localized abdominal discomfort followed



Heart dullness percussed out on chest colored red Purplish area on cheek indicates where sweating occurs white area absence of sweating Fingers of right hand show clubbing



Constricted pupil on right dilated pupil on left and absence of sweating on right indicate loss of sympathetic nerve connection



Angiocardiogram shows opaque dye in the left outflow tract of heart and in aneurysm of innominate artery

Case 73 Aneurysm of Innominate Artery

The diagnosis is aneurysm of the innominate artery due to syphilis

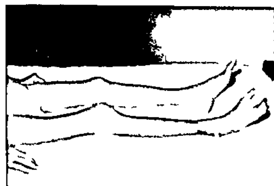
This 50 year old man had had a primary chancre on his penis 20 years before His blood and spinal fluid Wassermann were still positive Blood pressure was 210/92 A soft high pitched diastolic murmur was heard to the left of the sternum There was great enlargement of the left ventricle The right pupil was smaller than the left and sweating was absent on the right side of the face indicating loss of function of the sympathetic nerve fibers in this area The pulses and oscillometric readings in the right arm were very markedly reduced Clubbing of the fingers of the right hand developed

The upper left figure shows the area of the heart dullness percussed out on the chest colored with red The cheeks have been covered with starch paste and iodine The purplish area indicates where sweating occurs the white area absence of sweating Note that over the right side of the face sweating has not occurred Fingers of the right hand show marked clubbing

The upper right figure shows the constricted pupil on the right and the dilated pupil on the left and absence of sweating on the right The lack of sweating plus the dilated pupil are indicative of the loss of sympathetic nerve action

The lower figure is an angiocardiogram It shows the opaque dye in the left outflow tract of the heart and in the aneurysm of the innominate artery

Prognosis Without therapy this aneurysm could be expected to enlarge progressively Eventually it might rupture or erode through the adjacent structures such as the bronchus the esophagus the ribs or the spinal column In such cases antiluetic therapy must be given very cautiously to prevent an acute inflammatory reaction in the aneurysm Small doses of penicillin are indicated with a gradual increase in the size of the dose over a period of several months



Note atrophy of right leg



Large tumor formation over right lumbar and thoracic areas posteriorly



Aortogram showing anastomosis of vessels throughout pelvic lumbar and lower thoracic areas



Five years later Note increasing size of tumor

Case 74 Arteriovenous Aneurysm, Congenital

At the age of five this boy was found to have a lump in his right buttock The following year it was noticed that the right leg was warmer than the left leg A bruit was heard near the spine A diagnosis of arteriovenous anastomosis was made and the internal iliac and obturator arteries were ligated The bruit disappeared temporarily but returned a year later

This is characteristic of congenital arteriovenous anastomoses Usually there are many hundreds of tiny connections between the arteries and the veins One of these gradually increases in size until it becomes aneurysmal It gives rise to signs and symptoms which may be attacked surgically Removing the aneurysm then puts increased hemostatic pressure on one or more of the remaining anastomoses One or more of these in time usually develop into a large fistula For this reason multiple surgical procedures are often performed in such patients Surgery is usually not permanently successful but it may be advisable to attempt it in order to relieve temporarily the strain on the heart

The upper left photograph was taken when this man was 21 years of age Note the atrophy of the right leg which is located below the site of aneurysmal formation in the arterial tree The skin is tight and shiny The muscles are flabby and shrunken Edema is present over the foot which is larger than the opposite foot Pigment deposit is present up over the shins

The upper right photograph shows the large tumor formation over the right lumbar and thoracic areas posteriorly A large bruit is heard over this area Marked pulsation can be felt

The lower left figure represents an aortogram made by insertion of a catheter up the right femoral and iliac arteries into the abdominal aorta Contrast medium was then injected Note the great anastomosis of vessels present throughout the pelvis lumbar and lower thoracic area

The lower right photograph was taken five years later at the age of 26 Note the increasing size of the tumor Further surgery in this patient was considered to be impossible without damage to the renal arteries He is a permanent cripple with a poor life expectancy

Case 75 Arteriovenous Anastomosis







Dorsal view palmar view and close up of enlarged varix of fourth finger

Case 75 Arteriovenous Anastomosis, Congenital

This case is a 24 year old housewife From the age of two prominent veins were noticed in her right hand As she grew the right hand became larger than the left She also noticed that the temperature was warmer She was first seen by us at the age of 17 At that time a large pulsation was noticed in the palm of the hand A loud bruit was heard Surgery was performed and the arteriovenous anastomosis was excised Subsequently new anastomoses have made their appearance and several surgical procedures have been carried out but in this case as is usual in congenital arteriovenous anastomosis there are probably very many scattered small channels between arterioles and venules Under pressure these develop into large fistulas At present the patient has a loud continuous murmur at the wrist and another at the base of finger number five

The upper left photograph is a close up of the back of the hand The upper right photograph shows the palmar surface indicating dilated veins The lower photograph is a close up of the ring finger enlarged varix over which is heard a loud hum

Case 76 Arteriovenous Aneurysm



A



B



C



D



E

A Distended veins and hypertrophy of left leg B Note the large sack of veins A loud bruit as heard here C Aortogram showing "corkscrew" iliac artery D and E Five month follow p Notice disappearance of dilated veins and return of leg to normal size

Case 76 Arteriovenous Aneurysm Traumatic

Thirty years ago this 60 year old man was accidentally shot with a rifle bullet through the abdomen The bullet actually entered the right lower quadrant It pierced just above the bladder and came out in the left groin *In its course it nicked the left femoral artery* and vein An arteriovenous communication developed

Through the years massive distention of the veins occurred in the left leg which became swollen and heavy This excessive abnormal shunting of the blood into the systemic circulation which was of no physiologic use gradually put a strain on the heart Left ventricular hypertrophy followed and after many years decompensation occurred

The upper left figure (A) shows the greatly distended veins over the left thigh and leg Note the thickness of this leg as compared with the right leg

The next photograph (B) is a close up of the groin A large sac of veins is present here Over this area a large bruit is heard and a thrill is felt

The next photograph (C) is that of an aortogram Note the widely distended twisting artery

Surgery was performed during which this large arterial segment was removed and the veins draining into it were ligated Following surgery although no pulse was present in the femoral area of the groin strong popliteal and foot pulses were present indicating that excellent collateral flow was present

The next two photographs (D and E) were taken five months postoperatively Note the disappearance of the large dilated veins and the return of the leg to a normal size The close up of the groin shows the surgical incisions A small stitch abscess remains All the dilated veins have disappeared In contrast to the congenital arteriovenous fistula a traumatic fistula is apt to be single and when once successfully removed it will not return

6

Diseases of the Small Vessels*

INCREASED FRAGILITY OF VESSELS

1 Infectious purpura due to
  (a) Bacteria
  (b) Viruses and other micro organisms
2 Toxic purpura due to
  (a) Arsenic
  (b) Phosphorus
  (c) Phenolphthalein
  (d) Heparin and related substances
  (e) Coumarin derivatives and related substances
  (f) Venom
  (g) Other toxins
3 Purpura due to avitaminosis
  (a) Lack of vitamin C (scurvy)
  (b) Lack of vitamin K
  (c) Other vitamin deficiency
4 Purpura, secondary to increased venous pressure
5 Menstrual purpura
6 Senile purpura
7 Idiopathic purpura
  (a) Henoch's purpura
  (b) Schoenlein's purpura
8 Allergic purpura

INCREASED PERMEABILITY OF VESSELS

9 Urticaria and angioneurotic edema
10 Sensitivity to physical agents
  (a) Mechanical
  (b) Cold
  (c) Heat
11 Hematogenic purpura due to
  (a) Thrombocytopenia
  (b) Leukemia
  (c) Aplastic anemia
  (d) Granulocytopenia
  (e) Disturbances of clotting mechanism
12 Deficiency of blood clotting factors
13 Local inflammation
14 Anaphylactic shock
15 Traumatic shock
16 Burns
17 Frostbite

Not only the capillaries but also the finest arterioles and venules are included under the term small vessels

As the blood passes through the capillary network there is a constant interchange of certain of its constituents with the tissue fluids In conditions leading to increased pressure within the lumen or to damage to the capillary walls additional serum and formed elements may escape When there is an extravasation of cells into the skin or mucous membranes the term "petechiae" is used for

* Nomenclature and descriptions in this chapter are taken from the *Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Blood Vessels* 5th ed New York New York Heart Association Inc 1953

tumors or prolonged dependency of the legs as in long trips by air rail or automobile (especially while wearing constrictive garments such as girdles) Garments may be responsible for increased venous pressure with secondary hemorrhages Such purpuric areas are usually small in size and located about the feet and ankles They may also be found at the site of actual compression of the thighs

5 Menstrual Purpura This is a rare phenomenon of unknown cause It may occur in any part of the skin of the body except the face

6 Senile Purpura Purpura occurs in the elderly without adequate etiologic explanation although malnutrition must always be considered With the achlorhydria which is common to the aged the absorption of vitamin C may be interfered with

7 Idiopathic Purpura

(a) Henoch's Purpura This is a recurrent malady characterized by purpuric areas which may ulcerate The skin and mucous membranes are involved Attacks of acute abdominal pain may result from intestinal lesions

(b) Schoenlein's Purpura This is purpura with joint manifestations The diagnosis depends upon purpura appearing in the skin and associated with pains in the joints This may be due to hemorrhages in the joint capsules It is frequently classified as one of the rheumatoid group of diseases

8 Allergic Purpura This form of purpura occurs with or without thrombocytopenia

INCREASED PERMEABILITY OF VESSELS

Normally the capillary walls retain the solids of the blood and certain dissolved and suspended material of high molecular weight The capillary wall acts as an osmotic membrane allowing fluids and electrolytes to flow freely in accordance with physical laws Under certain conditions the capillary wall may be changed so as to allow protein and other constituents to flow through

9 Urticaria and Angioneurotic Edema This is a result of a localized capillary dilation and injury and edema of tissue spaces due to the action of histamine substances This may be secondary to a generalized sensitivity state or to local irritation as outlined below

10 Sensitivity to Physical Agents This results in local swelling due to extravasation of fluid into the tissue spaces of the skin which appears pale or red and may be due to

(a) Mechanical irritation (scratching pressure friction)

(b) Cold (allergic release of histamine)

(c) Heat (thermal damage to capillary walls)

11 Hematogenic Purpura This is not a primary disease of the minute blood vessels They are affected by certain blood conditions which cause purpura The defect in at least *some* of these conditions is a decrease of thrombocytes It is assumed that small openings are constantly being produced in the walls of capillaries due to trauma infection or loss of intercellular cement substance These holes are normally quickly plugged by blood platelets and fibrin with other cellular constituents If there is a defect in their number or chemotaxis the holes are not sufficiently plugged and hemorrhage ensues The commoner causative conditions are

minute hemorrhages 2 mm or less in diameter and which are often multiple and "purpura" for larger hemorrhages. Normally tiny cutaneous hemorrhages occur after even a brief constriction of a limb. These are probably due to increased capillary pressure plus the added factor of anoxemia of the cells of the capillary walls.

INCREASED FRAGILITY OF VESSELS

1 *Infectious Purpura* (a) Due to Bacteria. Petechiae are characteristic of *bacteremias and septicemias, notably in subacute* bacterial endocarditis. Any overwhelming bacterial infection may give rise to capillary damage which is reflected clinically by the appearance of hemorrhages in the skin and mucous membranes of pinpoint size or large enough to be called purpura.

(b) Due to Viruses and Other Microorganisms. Viral or rickettsial purpura is characteristic of chickenpox, smallpox and many viral infections. It is found nearly always in typhus and other forms of rickettsial diseases.

2 **Toxic Purpura** This may be caused by various poisons such as those listed below.

(a) Arsenic. Purpura may follow the administration of arsphenamine derivatives such as *Fowler's solution.*

(b) *Phosphorus*

(c) *Phenolphthalein. This drug is found* in many proprietary laxatives.

(d) Heparin. An overdose of heparin or heparin-like substances such as paritol and treburon will often cause purpura.

(e) Coumarin Derivatives and Vitamin K Blocking Agents. These substances include bishydroxycoumarin (dicumarol), ethyl biscoumacetate (tromexan), cyclocumarol, diphenadione and others. Slight trauma produces purpura in patients receiving significant amounts of anticoagulant drugs. In overdosage large hematomas may form.

(f) Venom. Certain types of snake and insect venom are toxic to capillaries (thrombogenic and hemolytic).

(g) Other Toxins.

3 *Purpura due to Avitaminosis*

(a) Lack of Vitamin C. Scurvy due to lack of vitamin C is characterized by easy bruising and bleeding gums from capillary oozing. Apparently the defect is in the loss of cement substance from the capillary walls themselves. Subclinical scurvy may be detectable only by the capillary fragility test and confirmed by finding a subnormal concentration of vitamin C in the blood. Careful examination of the skin about the ankles often reveals the first evidence of scurvy or of increased venous pressure. This physical finding is frequently missed and may be of real value. Specifically it is the thermolabile vi*tamin C which is lacking in scurvy.*

(b) Lack of Vitamin K. Lack of vitamin K produces a deficiency of prothrombin. This may result in hemorrhage from the capillaries.

(c) Other Vitamin Deficiency. It is claimed that vitamin P deficiency may cause capillary fragility but this is controversial.

4 **Purpura Secondary to Increased Venous Pressure** Increased venous pressure leads to an increase in capillary pressure. Small hemorrhages may result from rupture of these vessels. *Petechiae and purpura* are frequently seen as a result of defective venous *return* because of venous valvular defects, mechanical pressure on veins as from pelvic

Purpura of lips oral cavity and ala nasi



Hematoma and purpura Phlebitis in veins of lower leg





Large ecchymotic areas

thrombocytopenia myeloid or lymphatic leukemia aplastic anemia granulocytopenia and disturbances of the clotting mechanism such as hypoprothrombinemia Purpuric areas are also often seen associated with severe liver disease with associated disturbances in the coagulation factors in the blood

12 **Deficiency of Blood Clotting Factors** A deficiency of such factors as fibrinogen prothrombin Factor V Factor VII Factor VIII Factor IX Factor X and the Stuart Prower Factor while primarily due to defects in constituents rather than in vessel walls may result in easy bleeding into the skin These may be congenital hereditary or in some instances acquired A detailed discussion of them is beyond the scope of this volume

13 **Local Inflammation** A change in capillary permeability is characteristic of inflammation of all types

14 **Anaphylactic Shock** This may be associated with wheals or giant urticaria or edema

15 **Traumatic Shock** The picture of shock may be accompanied by the appearance of urticarial wheals sometimes blisters

16 **Burns** These may cause massive exudation and loss of fluid into the tissue spaces

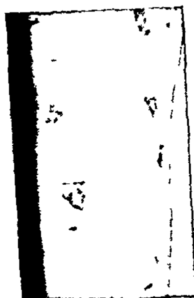
17 **Frostbite** This is discussed elsewhere

Case 77 Purpura with Agranulocytosis

This elderly woman complained of pains in her joints She consulted a physician who prescribed phenylbutazone 100 mg to be taken after each meal for a period of seven days The woman obtained a great deal of relief from this medication and decided to continue it without consulting her physician She kept up the drug for a period of six months Suddenly she developed bleeding in her mouth legs and thighs Examination disclosed that the bone marrow had ceased to produce granulocytic cells There was also a complete absence of platelets in peripheral blood smears Phlebitis had developed in the veins of the legs as well as large hematomas and widespread ecchymoses Bleeding had also occurred in the lips in the oral cavity and about the nose

The upper left illustration shows the purpura of the lips oral cavity and alae nasi In the upper right illustration hematoma and purpura are shown in the purple and dark red areas Phlebitis is present in the veins of the lower leg The lower illustration shows large ecchymotic areas

Case 79 Subclinical Scurvy



This 30 year old housewife had had very markedly increased vasomotor tone since childhood On exposure to cold blotches of purple discoloration occurred on the skin of the legs and arms The hands became cold pale and moist Tiny ulcers appeared Her diagnosis was livedo reticularis which has been discussed in another chapter An additional finding was present in her case which contributed to the formation of the ulcers She gave a history of never eating any fresh fruit salads or raw food of any kind with the exception of milk

A Rumpel Leede test was done using the Wright modification This consists of placing a blood pressure tourniquet around the arm in the usual manner pumping the pressure up to halfway between systolic and diastolic and leaving it in place for a period of 15 minutes At the end of that time the petechiae are counted in two circles each 1 inch in diameter The circles are located 2 inches below the elbow crease Normally there should be fewer than 10 per circle Ten to twenty is considered marginal and over 20 pathologic While this is characteristic it is not specific for scurvy In this patient an innumerable number of petechiae developed as shown in the illustration showing subclinical scurvy revealed by the capillary fragility test Her blood vitamin C level was 0 2 mg per 100 ml the normal is 0 8 to 1 2 mg She was given large doses of vitamin C orally and intravenously *namely*, 1000 mg per day by both routes The capillary fragility quickly returned to normal the ulcerations healed

Case 78 Senile Purpura



This 76 year old man complained of purplish spots on the skin of his legs A complete examination showed no positive findings that were not compatible with his advanced age The blood count was normal Bleeding time clotting time platelet count blood clot retraction prothrombin time capillary fragility tests were normal The patient was also examined from the point of view of a lesion in his prostate gland None was found Carcinoma of the prostate frequently metastasizes to the bone marrow and may give rise to purpura Carcinoma of the prostate also produces a proteolytic enzyme which sometimes gives rise to purpura and to hematuria In this patient the diagnosis of senile purpura is arrived at by the exclusion of other conditions such as defects in his blood clotting mechanism in his hematopoietic system or carcinoma of the prostate This is a common condition which is rarely a serious problem

The photograph shows senile purpura The fresh lesions are purple older ones are brown from hemosiderin formation

HYPERTENSIVE ISCHEMIC ULCERS

In some patients with arterial hypertensive disease the arterioles of the skin become sclerotic and thrombosed Small areas of skin may become hemorrhagic then necrotic This can occur even in the presence of strongly pulsating larger vessels

Case 81 Hypertensive Ischemic Ulcers



Large necrotic ulcer in lower third of tibia



Pinch grafts covering ulcer

This patient is a man 41 years of age He had been known to have had high blood pressure for many years His pressure while under our observation ranged from 200 to 250 systolic over 120 to 140 diastolic All the peripheral pulses were strong oscillometric readings were normal The left figure shows the large necrotic ulcer that had formed over the lower shin This was debrided and pinch grafts applied as shown in the right figure Successful healing was obtained The patient responded very well to Rauwolfia therapy His blood pressure fell to a range of 150/90 and has been maintained in this region for a four year period of follow up During this time no further ulceration has developed

Case 80 Clinical Scurvy



Coalescing petechial rash with extensive hemosiderin formation from old hemorrhage A small ulcer has formed



Wright modification of Rumpel Leede test

This 60 year old woman complained of a rash over her lower extremities She gave a history of having abstained from all raw foods for several years A Rumpel Leede test Wright modification as shown in the right figure gave innumerable petechiae Blood vitamin C level was 0.3 mg per 100 ml She was given 1000 mg per day of vitamin C orally and injections of a similar amount intravenously twice weekly The increased capillary fragility rapidly returned to normal The rash cleared except for the hemosiderin deposits which remained as a brown stain

The left figure shows a coalescing petechial rash Extensive hemosiderin formation from old hemorrhage is present A small ulcer has formed The right figure shows the Rumpel Leede test Wright modification Note the showers of petechiae below the level of the blood pressure cuff

Subclinical and even clinical scurvy are very common in our present civilization because of the frequency of diet faddists persons who believe all citrus fruits disagree with them In the treatment of peptic ulcers and colitis the physician should always ensure an adequate intake of vitamin C

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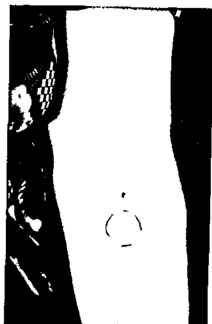
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7

Blood Vessel Tumors

HEMANGIOMA

A hemangioma (birthmark etc) is a benign growth often present at birth It consists of vascular spaces supported by a variable amount of fibrous stroma Hemangiomas may be subdivided into the following types

1 Cavernous Hemangioma This is a hemangioma which contains large vascular spaces This fact may be ascertained by the appearance of the lesion and especially by pressure upon the lesion It gives the sensation of a reduced resistance to pressure as compared to normal tissue

2 Capillary Hemangioma A hemangioma which contains small vascular spaces is a capillary hemangioma This appears as a small red spot Pressure produces a paling of the area

3 Plexiform Hemangioma A plexiform hemangioma is one which is not well circumscribed due to extensive ramification of the blood vessels that often penetrate deeply into the surrounding tissue

4 Sclerosing Hemangioma This is a hemangioma in which the blood vessels are constricted by proliferating fibrous tissue In the end stage many of the vessels are obliterated giving the lesion a superficial resemblance to a fibroma The sclerosing process is only evident clinically after long continued observation

5 Syndromes with Hemangiomas These are rare conditions

(a) Multiple hemangiomas and chondromas (Kast s syndrome)

(b) Hemangiomas of the retina and central nervous system sometimes associated with cysts of the pancreas and kidney (Lindau s or von Hippel Lindau s disease)

HEMANGIOENDOTHELIOMA

6 Benign

7 Malignant A hemangioendothelioma like a hemangioma contains vascular spaces but unlike a hemangioma it also contains solid nests of proliferating endothelium There are all gradations between lesions in which the proliferative endothelium is markedly anaplastic with the vascular spaces crudely formed to the opposite extreme in which the vascular spaces are prominent and the

Classification and description from the *Nomenclature and Criteria for Diagnosis of Diseases of the Heart and Blood Vessels* 5th ed New York New York Heart Association Inc 1953

hages manifested by epistaxis hematuria etc The disease is said to be transmitted by either sex as a simple Mendelian dominant

(b) Papillary Varices (Senile Angioma Cayenne Pepper Spot) These are small red compressible localized benign cutaneous swellings consisting of a single dilated vessel and occurring with increasing frequency after middle age They are benign

(c) Spider Angioma (Spider Nevus Nevus Araneus) This is a cutaneous lesion consisting of a central pulsating arteriole from which small dilated capillaries radiate outward producing a fanciful spider like appearance The lesions often are associated with liver diseases and pregnancy but may occur independently The area may vary from a few millimeters to several centimeters

proliferative endothelium less conspicuous It is impossible to differentiate morphologically between hemangioendotheliomas which metastasize and those which are locally invasive

Clinical manifestations depend upon the organ involved When the skin is involved there is a circumscribed tumor usually bluish and usually compressible It may be tender or painful or both The size varies from a few millimeters to several centimeters

SARCOMA

8 **Angiosarcoma** This term is seldom used Many pathologists reserve the term sarcoma for neoplasms arising from mesenchymal derivatives other than blood vessels If many of the neoplastic cells are anaplastic *but in places sufficiently differentiated to form* vascular spaces the pathologist makes the diagnosis of malignant hemangioendothelioma rather than angiosarcoma Some sarcomas are so highly undifferentiated that the pathologist cannot be certain of the parent tissue

9 **Kaposi s Sarcoma** This term is applied to multiple pigmented lesions composed of dilated capillaries fibroblastic proliferations inflammatory cells and hemosiderin deposits The hemosiderin deposition is due to extravasations of blood from ruptured capillaries

The lesions usually appear first in the skin of the extremities There is a bluish mass of blood vessels extending into the adjacent tissues and of slow growth Males are more commonly affected than females Autopsies have *demonstrated that visceral lesions do occur* but they are usually clinically undetectable The average life expectancy is said to be 5 to 10 years It is not at all certain whether this entity is really a sarcoma Some pathologists believe that it is essentially an inflammatory lesion

10 **Ewing s Sarcoma** It is widely held that Ewing s sarcoma is essentially an endothelioma and indeed the tendency of the neoplastic cells to surround spaces does suggest an endothelial origin However Ewing s sarcoma always arises in the skeletal system frequently in the vertebrae It is therefore listed under bone tumors The initial symptom usually is pain

11 **Glomus (Angioneuromyoma)** This benign tumor is a hypertrophied caricature *of the specialized* arteriovenous anastomoses found in the periphery and which are concerned with temperature control It consists of blood vessels imbedded in smooth muscle epithelioid cells and nonmyelinated nerve fibers These benign tumors may be situated anywhere in the skin but classically they are found on the hands and feet frequently beneath the nails Typically the tumor causes paroxysms of severe radiating pain especially on pressure upon the tumor The tumor is usually small seldom measuring more than a few millimeters in diameter and when superficial is slate gray in color Complete excision results in cure

12 **Hemangiopericytoma** This term is used to refer to a neoplasm which resembles a *glomus except that the epithelioid cells* are lacking Most are benign

13 **Telangiectasis** Telangiectasis may not be a true neoplasm It may be a dilatation of preexisting vascular channels either venules capillaries or arterioles rather than a neoplastic proliferation of new vessels

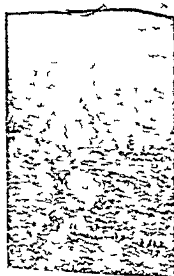
(a) Hereditary Hemorrhagic Telangiectasis This is characterized by numerous dilatations of the cutaneous and *mucous membrane* capillaries and venules These anomalies seldom measure over 4 mm Rupture of the mucous membrane lesions occasions hemorr



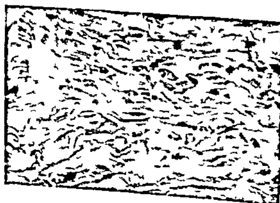
Multiple areas of tumor on leg



Lesions under tongue and on lips



Biopsy in low power



Biopsy in high power

Case 82 Kaposi s Sarcoma (Multiple Hemorrhagic Hemangioma of Kaposi)

This middle aged man noted peculiar brownish discoloration on his feet and shins five years previously The areas itched but otherwise did not bother him The lesions slowly progressed up the legs They were accompanied frequently by tiny hemorrhages into the skin The hemorrhages subsided and left behind deposits of hemosiderin Finally lesions appeared on the mucous membranes of his mouth and lips

The upper left figure shows multiple areas of tumor on the leg The area marked off by crayon was biopsied and is shown in the two lower figures *Note the mottled discoloration* caused by deposits of hemosiderin also the tiny darker areas of fresh hemorrhage

The upper right figure shows lesions under the tongue and on the lips The lower left figure is the biopsy in low power Note the deposits of hemosiderin the overgrowth of endothelial cells with a tendency to form new capillaries The lower right figure is a high power view of the lesion Note the sarcomatous changes in the vessels pyknotic nuclei and the numerous mitotic divisions

TREATMENT *The lesions can often be held* in check for many years by judicious use of x ray therapy

PROGNOSIS From the time of diagnosis the *life span is usually 5 to 10 years* Visceral lesions do occur that are often thought to be part of the primary tumor

Case 84 Kaposi s Sarcoma in a Middle Aged Woman



Lesions in both feet and toes



Ulcerated areas on internal malleolus

The left figure shows the lesions in both feet and toes Note the symmetry of the lesions the dark brown staining of hemosiderin and the intense brown color of fresh lesions

The right figure shows the internal malleolus Note the ulcerated areas where the tumor has eroded the skin also note the dark brown fresh deposits of tumor on the inner surface of the second toe

Case 83 Kaposi s Sarcoma



This 75 year old man developed this malig nant tumor of blood vessels at the age of 70 He had received extensive x ray therapy This slowed up the progress of his disease which advanced slowly but definitely The photo graph shows the tumor infiltration in the feet legs and hand The brown areas are sites of hemosiderin deposits from hemorrhages in the tumor tissue A small ulcer over the dorsum of the foot has appeared

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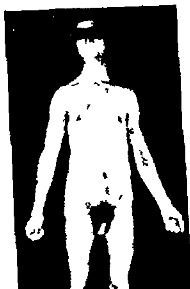
The right figure shows the internal malleolus Note the ulcerated areas where the tumor has eroded the skin also note the dark brown fresh deposits of tumor on the inner surface of the second toe

Case 85 Angiosarcoma



The illustration is the leg of a 52 year old woman who developed a purplish lesion on her foot one and a half years previously The lesion gradually spread to involve a wide area Note the extensive infiltration over the ankle above the instep as well as the patchy infiltration in the midleg Biopsy of the lesion showed a malignant sarcoma of blood vessel type Metastasis had developed in the groin The case was too late for surgical intervention X ray therapy was used for palliation

Case 86 Cavernous Hemangioma



Extensive lesions on fore arm arm thorax chest and abdomen



Close up of large cavernous areas on the arm

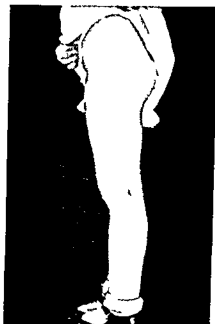
This 30 year old man was born with extensive cavernous hemangiomas of his trunk and arms Seven years ago at age 23 he commenced to have Jacksonian epileptic seizures probably indicating a similar lesion in the cortex of the brain

The left figure shows the extensive lesions on the forearm arm thorax chest and abdomen The right figure is a close up showing the large cavernous spaces on the arm Because of the extensive degree of the lesions in this patient no therapy was attempted

Case 87 Cavernous Hemangioma



Dilated pools of veins merging into large hemangioma



Distribution of lesion from knee to hip area

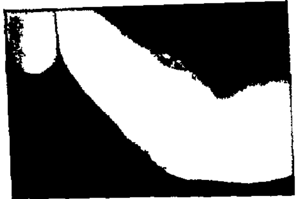
This 10 year old girl was born with an extensive hemangioma

The left photograph shows dilated pools of veins which merge into a large hemangioma No signs of arteriovenous anastomosis could be demonstrated The right photograph is a lateral view showing the distribution of the lesion from the knee to the hip area At a younger age this could have been treated with sclerosing injections with very little if any scarring It might have been quite successful At this age because of the wide expanse of the lesion whatever type of therapy is attempted will leave scarring

Case 88 Cavernous Hemangioma



Before therapy



One year follow up

This two week old baby has a large cavernous hemangioma in the elbow region (left figure) Many of these lesions decrease in size as the child grows but this covered such a large area and involved such large vessels that it was deemed advisable to treat it at once The lesion responded quickly to sclerosing injections using Sylnasol Very little scarring resulted A series of 10 injections at one month intervals was given with the result shown in the right figure one year after therapy

Case 89 Cavernous Hemangioma



Fungating tumor



Two month follow up

This six month old infant has a large fungating cavernous hemangioma which extrudes above the surface and penetrates deep into the tissues of the back She was treated by implanting radon seeds into the tumor mass The left illustration shows the extensive tumor The right illustration was taken two months later Notice that the tumor has shrunk the fungating area has sloughed and been covered by epithelium Small telangiectases remain around the periphery When the child is about eight or nine years of age plastic surgery will be done removing the scarred area and pulling the tissues together to give a good cosmetic result

Case 90 Capillary Hemangioma



Capillary hemangioma on face



Three year follow up

This child was born with a large red capillary hemangioma on the face corresponding to the third or submaxillary division of the trigeminal nerve It covered the cheek as shown in the left illustration extending from the jaw line to the ear and across the face Because these lesions frequently fade as the child grows older and because whatever form of treatment used would leave some scar formation it was deemed advisable to watch this patient and to postpone therapy

The second picture at the right was taken three years later The lesion has shrunk in size and in color and is hardly visible except when the patient cries as in the photograph As times goes on it can be expected to fade still further In later life it will be possible to conceal the residual traces of the lesion with cosmetics Some of these capillary hemangiomas persist into adult life at which time satisfactory cosmetic result can *sometimes* be obtained by tattooing them so that they blend in with the normal color of the skin This type of tattooing has been described by Herbert Conway (Tumors of the Skin Springfield, Ill Charles C Thomas and Co 1956)

Case 91 Capillary Lymphangioma



Capillary hemangioma covering left side of body



Close up of leg showing lesion and ulcer above malleolus

This 35 year old colored woman has a capillary lymphangioma which covers almost the entire left half of her body Notice how the lesion shown in the left figure involves the half of the face the left arm the left chest and the left leg

The right figure is a close up of the leg illustrating the diffuse nature of the lesion and the formation of an ulcer above the malleolus This tissue is very fragile and easily traumatized When ulcers form they are difficult to heal In this case healing of the ulcer was induced by elevation rest and antimicrobial ointments The diffuse primary lymphangioma could not be satisfactorily treated

Case 92 Pulsating Angioma



Close up of lesion on forehead



Two lesions on elbow

This is also known as primary angioma nevus araneus or spider nevus

This 27 year old girl noticed the development of small red skin lesions since the menarche They are located on the arms trunk legs with several on the face Each is composed of a central red dot which pulsates and from which small vessels radiate like the spokes of a wheel There is no family history in her case nor has there been any spontaneous bleeding from the nose or other mucous membranes such as that characteristic of the hereditary Rundu Osler Weber syndrome She is in excellent health her liver and endocrine status is normal

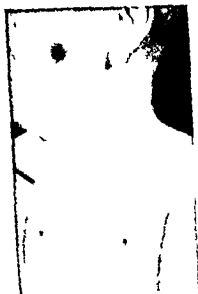
The left illustration shows a close up of the lesion on her forehead The right illustration shows two of the lesions on her elbow When these lesions occur in other patients in the mucous membranes they are apt to bleed profusely and extensively on slight trauma Recently it has been suggested that the lesions are associated with an imbalance of estrogen production or estrogen conjugation by the liver Therapy has been directed to correcting this factor with success in some cases

Case 93 Hematocele



This four year old boy fell off his tricycle and traumatized his left supraclavicular area. A large round swelling shown in the photograph developed in the neck. It simulated a cyst or a lymphocele from trauma to the lymphatic duct. Aspiration proved it to be an hematocele. It subsided without further therapy. The prompt application of ice to developing hematoma is of great value in stopping the formation of these lesions.

Case 94 Pulsating Angioma



In this patient (The New York Hospital #40-066) numerous large spider angiomas were found on the cheek at the angle of the jaw the region of the clavicle and over the sternum as seen in the illustration These lesions are frequently found in patients who have portal hypertension due to cirrhosis of the liver They are most commonly found externally at the level of the diaphragm At present they are believed to be due to a loss of the liver's ability to conjugate estrogenic hormones and are a sign of decreased liver function This however is not conclusively established as the definitive explanation This 57 year old patient died of cirrhosis of the liver

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